

# Pseudoexfoliation syndrome, ocular and systemic associations.

Dr. Haider Aswad. Al-Hemidawi, CABOphth FRCS. FICO/Al-Qadisiya University.

Dr. Saif Abbas. Al-Shamarti, CABOphth. FRCS. JMC .ICO/Al-Qadisiya University.

Dr. Hasenen Hasan. Al-Rekabi, FICMS. FICO/Al-Qadisiya University.

## Abstract

**Background:** Pseudoexfoliation syndrome (PXF) is a recognized risk factor for developing cataract, glaucoma and lens dislocation. PXF also associated with increased risk of vascular disorders and hearing loss, this study aims to assess the prevalence of this syndrome among patients (>40years) attend ophthalmology clinic in Diwaniyah teaching hospital with emphasis to both ocular and systemic associations.

## Methodology:

2680 patients age more than 40 years who attend the ophthalmology clinic in Diwaniyah teaching hospital for the period from July 2013 to January 2015 were recruited by the researchers, detailed systemic and ocular history are taken, detailed ophthalmological examination done including visual acuity testing, refraction, slit lamp biomicroscope examination, applanation tonometer and gonioscopy, dilated examination of the lens using

the LOCS II grading system for cataract and fundus with optic nerve examination.

All the patients have been sent for the ENT department for hearing examination and assessment, also send for the internal medicine department for detailed cardiovascular examination and risk assessment.

## Results:

Out of 2680 patients who enrolled in this study, 216 had PXF so the percentage of PXF will be 8% in patients above 40, Mean age of the patients is  $65.43 \pm 13.94$  years, range 41 to 93 years, PXF is associated with increased incidence of cataract (50,46%), glaucoma (8,9%), hearing loss (11,57%), Hypertension (36,57%) and DM type II (38,69%).

**Conclusion:** PXF appear to be a common disease in Iraqi population, PXF is significantly associated with increased incidence of cataract,

glaucoma, hearing loss, Hypertension and DM type

II.

## Introduction

The pseudo exfoliation syndrome(PXF) sometime known as exfoliation syndrome, is a relatively common cause of chronic open angle glaucoma, though subtle signs are easily overlooked, when an eye with PXF develop glaucoma the condition is known as pseudo exfoliation glaucoma PXG<sup>1</sup>

Pseudo exfoliation syndrome PXF has been first described in 1917 by Linberg in Finnish population<sup>2</sup>, the clinical diagnosis is made by the presence of typical pseudo exfoliation materials (PXM)on the anterior capsule surface , in addition other features include endothelial pigmentation loss of pupillary ruff ,iris trans illumination ,Sampolisi line, and pigment deposition in the trabecular meshwork<sup>3</sup>,PXF is associated with various ocular complications ,elevated intraocular pressure and glaucomatous nerve damage had been demonstrated in patients with PXF.<sup>4-8</sup>

Cataract were reported to be more common in patients with PXF<sup>9,10</sup>, Cataract surgery is more

hazardous due to combination of poorly dilating pupil, increased risk of zonular dialysis and capsular tear. Other problems include a postoperative pressure spike, corneal oedema, increased incidence of capsular opacification and contraction, and IOL subluxation<sup>1</sup>.

PXF is considered to be a systemic disorder PXM has been reported in lungs, skin, liver, heart, Kidney, gallbladder, blood vessels, extra ocular association between amuscles and meninges<sup>11</sup>, the PXF and sensorineural deafness has been reported<sup>12</sup>.

PXF is rarely seen before the age of 40 and its prevalence increase markedly with age<sup>13</sup>, although it occurs virtually in in every area of the world, a considerable racial variation exists, prevalence rate ranging from 0% in Greenland Eskimo to 21% in Icelanders<sup>14</sup>

The aim of this hospital based study is to estimate the prevalence of PXF and provide a descriptive analysis whether this syndrome associated with increased incidence of cataract and glaucoma, hearing loss, DM hypertension and other cardiovascular diseases.

## Methods

Patients above 40 years in age who attended eye clinics in Diwaniya teaching hospital for the period from July 2013 to January 2015 were invited to participate in this study; detailed

medical and ocular history is taken including history of DM, hypertension, hearing loss, visual problems, corrective glasses, previous surgical history.

Complete ocular examination was done including visual acuity testing with Snellen chart, refraction with correction, slit lamp biomicroscope examination using Haag- Streit Slit lamp for the anterior segment, applanation tonometer using Goldman tonometry and gonioscopy with Goldman three mirror, dilated examination of the lens using the LOCS II grading system for cataract and fundus with optic nerve examination.

PXF is diagnosed clinically by the presence of PXM on the lens surface or at the pupillary border or on trabecular meshwork on gonioscopy examination with/out Sampolisi line and pigment deposition on the angle and /or corneal endothelium.

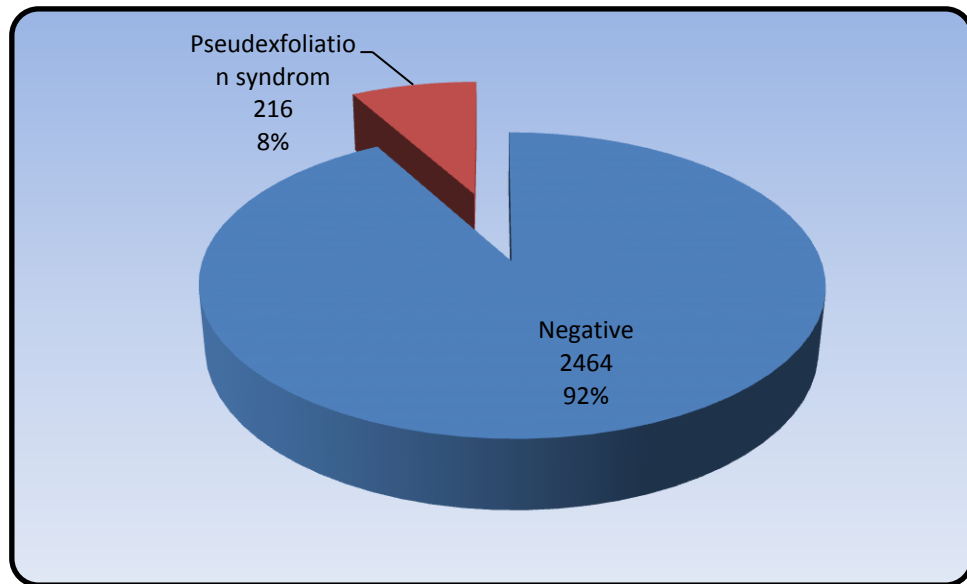
Cataract are graded on slit lamp using LOCSIII grading system for cataract, lens opacities were defined as no cataract ( $avLOCS < 1.5$ ) and cataract ( $avLOCS > 1.5$ ).

Diagnosis of glaucoma is made using the International society of Geographical and Epidemiological ophthalmology classification (ISGEO) if IOP  $> 22$ mmHG in either eye with vertical C/D ratio  $> 0.7$  or difference in vertical C/D ratio  $> 0.2$  with focal thinning, vertical notching, or splinter hemorrhage.

All the patients have been sent for the ENT department for hearing examination and assessment, also send for the internal medicine department for detailed cardiovascular examination and risk assessment.

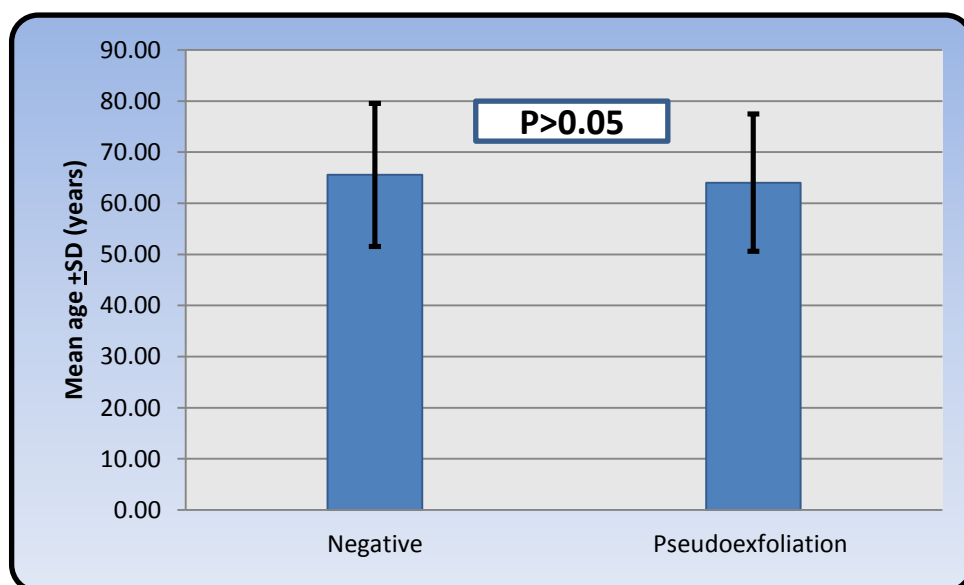
## Results

Out of 2680 patients who were enrolled in the present study, 216 had PXF so the percentage of PXF will be 8% in patients above 40 in Iraq as in the pie chart in figure “1”.



**Figure 1: show percentage of PXF in patients above 40 years**

There was no significant difference in mean age of patients with PXF than those without PXF ( $65.56 \pm 13.98$  versus  $64.02 \pm 13.44$ );  $P > 0.05$ , as shown in figure “2”.



**Figure 2: show mean age of patients with PXF versus patients without PXF**

Unilateral PXE was noted in 23.45 % of the patients while bilateral was found in 76.55%. Out of the 216 patient with PXF, 99 were male and 117 were female, the association between gender and PXF was not statistically significant; **P>0.05.**

Cataract was significantly more frequent in patients with PXF than in patients without, (50.46%) versus (14.29%); **P<0.001.** "Table 1".

**Table 1: show incidence of cataract in patients with PXF versus patients without PXF**

Cataract	Pseudoexfoliation					
	Yes		No		Total	
	No.	%	No.	%	No.	%
Yes	109	50.46	352	14.29	461	17.20
No	107	49.54	2112	85.71	2219	82.80
Total	216	100.00	2464	100.00	2680	100.00

**P<0.001**

Glaucoma was significantly more frequent in patients with PXF than in patients without, (8,9%) versus (1,3%); **P<0.001.** "Table 2".

**Table 2: show incidence of glaucoma in patients with PXF versus patients without PXF**

Glaucoma	Pseudoexfoliation					
	Yes		No		Total	
	No.	%	No.	%	No.	%
Yes	52	24.07	186	7.55	238	8.88
No	164	75.93	2278	92.45	2442	91.12
Total	216	100.00	2464	100.00	2680	100.00

**P<0.001**

Hearing loss was significantly more frequent in patients with PXF than in patients without, (11, 57%) versus (1, 18%); **P<0.001** as shown in "Table 3".

Table 3: show incidence of Hearing loss in patients with PXF versus patients without PXF

Hearing loss	Pseudoexfoliation					
	Yes		No		Total	
	No.	%	No.	%	No.	%
Yes	25	11.57	29	1.18	54	2.01
No	191	88.43	2435	98.82	2626	97.99
Total	216	100.00	2464	100.00	2680	100.00

**P<0.001**

Hypertension was significantly more frequent in patients with PXF than in patients without, (36, 57%) versus (7, 71%); **P<0.001** as shown in "Table 4".

Table 4: show incidence of Hypertension in patients with PXF versus patients without PXF

Hypertension	Pseudoexfoliation					
	Yes		No		Total	
	No.	%	No.	%	No.	%
Yes	79	36.57	190	7.71	269	10.04
No	137	63.43	2274	92.29	2411	89.96
Total	216	100.00	2464	100.00	2680	100.00

**P<0.001**

Diabetes mellitus type II was significantly more frequent in patients with PXF than in patients without, (38, 89%) versus (15, 3%); **P<0.001** as shown in "Table 5".

Table 5: show incidence of DM II in patients with PXF versus patients without PXF.

Diabetes mellitus	Pseudoexfoliation					
	Yes		No		Total	
	No.	%	No.	%	No.	%
Yes	84	38.89	377	15.30	461	17.20
No	132	61.11	2087	84.70	2219	82.80
Total	216	100.00	2464	100.00	2680	100.00

**P<0.001**

## Discussion

The reported rate of PXF syndrome in different population show extensive variation , it could be low as in Eskimo 0% <sup>1</sup>, or high as in Navajo Indians 38% <sup>2</sup>, while In neighboring countries the estimated rate was as follow: in study done in Jordon the rate was 9.1% <sup>3</sup>, in study done in Egypt it was 4.14% <sup>4</sup>, Iran 6.9% <sup>5</sup>, Saudi Arabia 9.3% <sup>6</sup>, this variation in prevalence could be due to racial differences ; population based and hospital based studies ; persons over certain ages could affect the results of the study.

One of the source of bias in this study that it was a hospital based study since PXE is silent disease could be not discovered until the patient consult his doctor, so over or under estimation of the prevalence could be attributed to its hospital based nature of the study.

Our patients are recruited form the eye clinics in Diwaniya teaching hospital, the prevalence between males (99) and females (117) show no statistically significant difference, the study show that bilateral disease is much more prevalent (76.55%.) than unilateral one (23.45 %).

The study also show increase incidence of cataract ( about three folds more )in patients with PXF than normal , and about three folds more incidence of glaucoma in patients with PXF than normal ,a strong relation between PXF and glaucoma is known <sup>17</sup>,patient with PXF has two to three folds increase incidence of glaucoma according to the Blue mountain eye study 26,while other studies demonstrate that eyes with PXF has higher mean IOP <sup>15, 16</sup> ,moreover Topouzis F et al .report an increase in the likelihood of glaucoma at the same IOP in patients with PXF than others<sup>18</sup>,our study is consistent with these study regarding the strong association of PXF with glaucoma ; Moreover the rate is higher than the rate in other similar studies like the Blue mountain eye study ( 14.2%) this over estimation is one of the limitations of hospital based study.

A significant association also found regarding systemic complications usually associated with PXF, regarding hearing loss; the study shows more prevalent in patients with PXF than normal ;(11,57%) in PXF while it found in (1,18%) in normal, hypertension also more common with PXE (36,57%) while normal (7,71%): DM type II in PXF also more

common (38,89%) than patient without PXF (15,3%).

## Conclusion & Recommendations

PXF is a common disease in Iraqi population is 8% according to this study, there is a strong association between PXF and cataract ;glaucoma should the ophthalmologist be aware of it, Moreover there is strong association between PXF and some systemic diseases like HT,DM type II and hearing loss which might suggest that PXF is a systemic disease affect the eye and other organs, the ophthalmologist should send all patients with PXF syndrome for full systemic assessment especially for the internal medicine department .and ENT department



## REFERENCES

- 1- Kaniski JJ, clinical ophthalmology. 2010; 355.
- 2- Lindberg JG. Kliniska under sökningar över depigmentering av pupillarranden genomlysbarheten av iris vid fall av aldersstarrsamt i normala ögon hos gamlapersoner (thesis) Helsinki, Finland. Helsinki University 1917.
- 3- Prince AM, Ritch R. clinical signs of the pseudoexfoliation syndrome. Ophthalmology 1986; 93:803-7.
- 4- Fukisawa K, Sugai S, Inomata H, et al. Relationship between intraocular pressure and age in exfoliation syndrome. Ophthalmologica 1995; 209:199-202.
- 5- Davanger M, Ringvold A, Blika S. Pseudoexfoliation, IOP, and Glaucoma, Acta Ophthalmol 1991; 69:569-73.
- 6- Kjørsvik S, Rossvold I. The middle Norway eye screening II. Prevalence of simple and capsule glaucoma. Acta ophthalmol 1991; 69:273-80.
- 7- Ringvold A, Blika S, Elsas T, et al. Increase occurrence of exfoliation in the male, Spanish American population of the New Mexico. J Am Optom Assoc 1992; 63:643-8.
- 8- Konstas AGP, Jay JL, Marshall GE, et al. Prevalence, diagnostic features, and response to trabeculectomy in exfoliation syndrome. Ophthalmology. 1993; 100:619-27.
- 9- Hirvelä H, Luukinen H, Laatikainen L. Prevalence and risk factors of lens opacities in the elderly in Finland. ophthalmology 1995; 102:108-17.
- 10- Rouhiainen H, Terasvirta M. Presence of pseudoexfoliation on clear and opacified lens in an aged population. Ophthalmologica 1992; 204:67-70.
- 11- Schlotzer-Schrehardt UM, Koca MR, Nouman GO, Volkholz H. Pseudo exfoliation syndrome, Ocular manifestation of a systemic disorder? Arch Ophthalmol 1992; 110(12):1752-6.
- 12- Cahill M, Early A, Stack S, Balyne AW, Eustace P. Pseudoexfoliation and sensorineural hearing loss. Eye (London) 2002; 16(3):261-6.
- 13- Aasved H. Mass screening for fibrillogenesis epitheliocapsularis, so called senile exfoliation or pseudo exfoliation of the anterior lens capsule. Acta Ophthalmol. (Copenh) 1971; 49(2):334-43.
- 14- Forsius H. Prevalence of PXF of the lens in Finns, Lapps, Icelanders, Eskimos and Russians. Trans Ophthalmol. Soc. UK. 1979; 99:296-298.
- 15- Hiller R, Sperduto RD, Kreiger DE. Pseudoexfoliation, IOP, and senile changes in a population based survey, Arch Ophthalmol, 1982; 110:1080-2.

- 16- Mitchell P, Wang JJ, Hourihan F, the relationship between glaucoma and pseudoexfoliation, The Blue mountain eye study. , Arch Ophthalmol 1999; 117:1319-24.
- 17- Henry JC, Krupin T, Schmitt M, Long term follow up of pseudoexfoliation and the development of elevated intraocular pressure. Ophthalmology 1987; 94; 545-52.
- 18- Topouzis F, Harris A, Wilson MR, Increased likelihood of glaucoma at the same screening intraocular pressure in subject with Pseudoexfoliation Am J Ophthalmol. 2009; 148; 606-613.

## الخلاصة

**الغرض من الدراسة:** - متلازمة النقشر الكاذب (PXF) هي عامل خطورة مهم لكثير من امراض العين كالماء الابيض والاسود وخلع العدسة، ايضا هي عامل خطورة مهم لكثير من امراض الجسم كأمرض القلب وتصلب الشرايين.

الهدف من هذه الدراسة هو معرفه مدى انتشار هذه المتلازمة وعلاقتها بأمراض العيون والجسم الاخرى.

**طريقه العمل:** - تم اختيار ٢٦٨٠ مريض بعمر أكثر من ٤٠ عام بصوره عشوائية من خلال مراجعي شعبه العيون في مستشفى الديوانية التعليمي للفترة من تموز ٢٠١٣ الى كانون الثاني ٢٠١٥، تم اخذ التاريخ المرضي المفصل للمريض وتم فحصهم بصوره كامله في شعبه العيون لكشف امراض العيون الموجودة ومدى تعلقها بمتلازمة النقشر الكاذب كما تم ارسال المرضى الى شعبه الانف والاذن والحنجرة لفحص السمع وتم ارسال المرضى ايضا الى شعبه الطب الباطني للفحص السريري الكامل.

**النتائج:** - ٢١٦ مريض مصاب بمتلازمة النقشر الكاذب من أصل ٢٦٨٠ بنسبه ٨٪، معدل عمر المرضى ٦٥\_١٣ سنه وتم الوصول الى النتائج الاتية: - متلازمة النقشر الكاذب (PXF) مرتبطة بزيادة نسبه حصول الماء الابيض (٤٦,٥٠٪) زيادة نسبه داء الزرقاء (٨,٩٪) وضعف السمع (١١,٥٧٪) وارتفاع ضغط الدم بنسبه (٣٦,٥٧٪) وداء السكري من النوع الثاني (٣٨,٦٩٪).

**الاستنتاجات والتوصيات:** - متلازمة التقشر الكاذب (PXF) هي مرض شائع في العراق وهي مرتبطة بزيادة

احتمالية حصول بعض الامراض كما ذكر انفا لذلك يوصي الباحثون بضرورة فحص المريض المصاب بهذا

المرض بصورة كاملة ودقيقه ومن قبل فريق من الاطباء.