

Association between Pseudoexfoliation Syndrome and Sensorineural Hearing Loss



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There are no conflicts of interest.

Pseudoexfoliation syndrome (PXF)

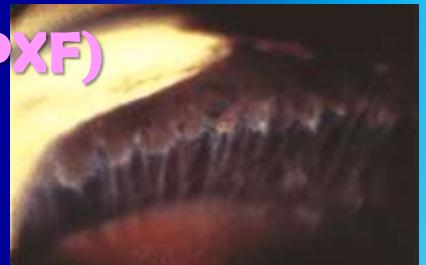
- Systemic disorder of the extracellular matrix affecting the eye and visceral organs
- It is an age-related disease.
- Abnormal fibrillar extracellular material is produced and accumulates in many ocular tissues.
- Lysyl-oxidase-like-1 (LOXL1) gene on chromosome 15q24 with abnormal elastin metabolism is implicated in PXF

Pseudoexfoliation syndrome (PXF)

- The fibrillar material can be found on all anterior segment structures bathed by aqueous humor.
- It appears as a 'dandruff-like' material on the anterior lens capsule deposited in a double concentric ring pattern.

Ritch R. Exfoliation syndrome. In: Ritch R, Shields MB, Krupin E, eds. *The glaucomas*. St. Louis, Mosby, 1996

Alward WLM: *Color atlas of gonioscopy*, San Francisco, 2000, Foundation of American Academy of Ophthalmology.

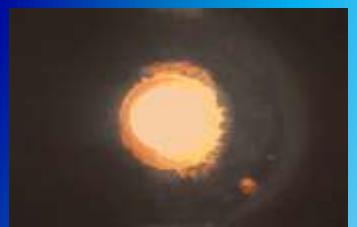


Pseudoexfoliation syndrome (PXF)

- Ocular manifestations include iris depigmentation, trabecular hyperpigmentation, 2nd glaucoma, poor pupillary dilatation, increased vessels permeability and phacodonesis.
- Pseudoexfoliation material has been identified in the conjunctiva, extraocular muscles, retinal, ciliary and vortex vessels.

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Pseudoexfoliation syndrome (PXF)

- Glaucoma associated with PXF is multifactorial.
- In the Blue Mountain study, eyes with PXF had five-fold increased risk for OAG. PXF is most common “identifiable” cause of open-angle glaucoma.
- Angle-closure glaucoma is also more frequent with PXF due to zonular weakness with anterior movement of the lens causing ocludable angle.
- Malignant glaucoma may be associated with a pseudoexfoliation

Pseudoexfoliation syndrome (PXF)

- Pseudoexfoliation material has been identified in the lung, liver, kidney, gall bladder, skin and cerebral meninges.
- Cardiovascular and cerebrovascular diseases such as angina, myocardial infarction, aortic aneurysm, hypertension and Alzheimer's dementia have been linked to PXF.
- Recently, PXF was found to be more frequent in women with pelvic organ prolapse.

Pseudoexfoliation syndrome (PXF)

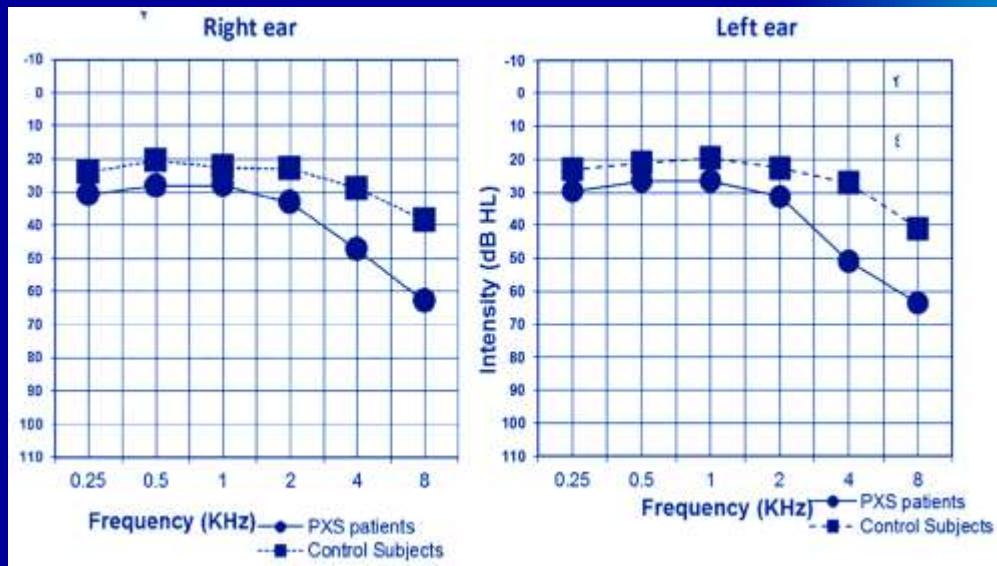
- Sensory-neural (SNHL) hearing loss, has been linked to PXF.
- SNHL in PXF syndrome may be caused by deposition of its material on the hair cells in the inner ear.

Association between Pseudoexfoliation Syndrome and Sensorineural Hearing Loss

- In a cross-sectional comparative study, 25 patients with PXF and 15 age and sex-matched normal controls were subjected to complete ophthalmological and audiological evaluation.
- The degree of sensory neural hearing loss (SNHL) was compared between the two groups and correlated with the clinical findings including glaucoma, cataract, and laterality of PXF.

- The patients were 13 females and 12 males with a mean age of 68.8 ± 7.9 years.
- Six patients had unilateral PXF and 19 patients had bilateral disease, with the total number of affected eyes was 44.
- Twenty five eyes (57%) had glaucoma and 38 eyes (86%) had cataract. There was a statistically significant association between PXF and each of glaucoma ($P < 0.001$) and cataract ($P < 0.001$) (related-sample McNemar test).

Mean hearing sensitivity of PXF patients and controls for the frequency range 250–8000 Hz.



Hearing sensitivity in the six tested frequencies in PXF patients and controls

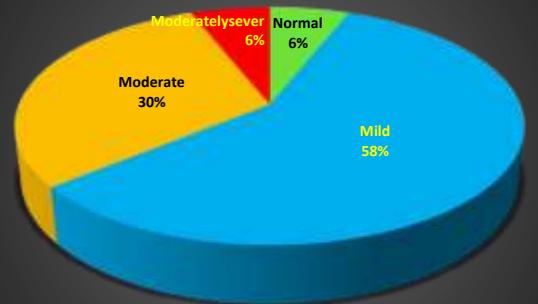
Frequency	Pseudoxfoliation patients	Control subjects	P value
250 Hz	30.2 ± 11.4	23.5 ± 2.5	0.035
500 Hz	27.3 ± 10.2	20.7 ± 3.3	0.001
1000 Hz	27.2 ± 11.1	21.1 ± 3.3	0.02
2000 Hz	32.2 ± 15.1	22.7 ± 5.4	0.048
4000 Hz	49.1 ± 21	28.0 ± 9.3	0.000
8000 Hz	63.3 ± 27	39.7 ± 14.8	0.001

Significantly worse hearing sensitivity (i.e. more degree of hearing loss) in PXF patients compared with controls (Independent samples of Mann Whitney U test)

Hearing Loss in Controls



Hearing Loss in PXF



Hearing sensitivity in PXF patients with glaucoma and PXF patients without glaucoma

Frequency	Glaucoma	No Glaucoma	P value
250 Hz	29.6 ± 10.9 dB HL	30.8 ± 12.2 dB HL	0.97
500 Hz	26 ± 8 dB HL	28.6 ± 12 dB HL	0.62
1000 Hz	28.2 ± 12 dB HL	26.2 ± 10.3 dB HL	0.56
2000 Hz	35.8 ± 17.3 dB HL	28.6 ± 11.9 dB HL	0.12
4000 Hz	52.6 ± 25.5 dB HL	45.6 ± 12.3 dB HL	0.31
8000 Hz	68.2 ± 31 dB HL	54.4 ± 23 dB HL	0.08

No statistically significant difference

(Independent samples of Mann Whitney U test)

Hearing sensitivity in PXF patients with cataract and PXF patients without cataract

Frequency	Cataract	No cataract	P value
250 Hz	30 ± 11.8	31.4 ± 9.4	0.38
500 Hz	27.4 ± 10.5	26.4 ± 8.5	0.98
1000 Hz	27.7 ± 11.5	23.5 ± 8.5	0.43
2000 Hz	33.1 ± 15.6	26.4 ± 10.6	0.4
4000 Hz	50.1 ± 20.5	42.8 ± 24.8	0.25
8000 Hz	63.6 ± 26	61.4 ± 31.4	0.76

No statistically significant difference

(Independent samples of Mann Whitney U test)

- In patients with unilateral PXF, comparing the ears towards the affected side with those towards the non-affected eye, across frequencies, the mean hearing sensitivity was 5 dB HL or less better in the ears towards the non-affected side.
- However, such difference was not statistically significant.

On comparing hearing sensitivity in patients with unilateral to those with bilateral PXF, no statistically significant difference was found at any frequency.

- Surgical difficulties in PXF include **weakness of zonular apparatus, reduced pupillary dilatation** and coexistence of glaucoma.
- Postoperative fibrosis with subsequent shrinkage of the capsule is increased in these eyes, and these centripetal forces will further loosen the zonular fibers.
- Late in-the-bag IOL dislocation is therefore anticipated in these patients.

- Hearing loss in PXF patients may add to difficulties during ophthalmic surgery when performed under topical or local anesthesia.
- This is due to the difficulty in communication with the patients not using hearing aids or those asked to remove their hearing aids in the operating room.

