# Unilateral Pseudoexfoliation in Two Young Patients with History of Iris Trauma and Associated Intraocular Surgery

*Ghasem Fakhraie, MD[1] • Massood Mohammadi, MD[2]*
*Golshan Latifi, MD[3] • Sasan Moghimi, MD[1]*

**Abstract**

***Purpose*:** To report two young patients with unilateral pseudoexfoliation (PEX) syndrome

***Case reports*:** Two young patients with PEX syndrome were examined. The first was a 30-year-old woman who had a history of penetrating ocular trauma with iris prolapse in the right eye that subsequently developed PEX material deposition on pupillary border and anterior lens capsule. The second was a 13-year-old girl with history of trabeculectomy in infancy and subsequent needle bleb revision in her right eye with PEX material deposits on anterior lens capsule. In both patients history of iris manipulation in the form of iridectomy was present. The time between ocular trauma or surgery with iris manipulation and detection of PEX syndrome were 16 and 11 years, respectively.

***Conclusion*:** According to a few previous reports of occurrence of PEX syndrome in young patients and surgical histories of our patients, there may be a role for ocular and particularly iris trauma in the development of PEX syndrome in younger patients.

***Keywords*:** Pseudoexfoliation Syndrome, Young Age, Iris Trauma, Iris Surgery

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## Introduction

Pseudoexfoliation (PEX) syndrome is a generalized disorder of extracellular matrix and its clinical manifestation is limited to ocular tissues. It is diagnosed by biomicroscopic finding of abnormal white flaky deposits on ocular structures that line the aqueous-bathed surfaces of the anterior segment.[1]

PEX syndrome and associated glaucoma are usually diagnosed in older patients and their prevalence increases with age.[1,2] There are few case reports of its occurrence in young patients and all reported cases had a history of previous ocular surgery or trauma.[2-6]

1. Associate Professor of Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran
2. Assistant Professor of Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran
3. Resident in Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran

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Correspondence to: *Massood Mohammadi, MD*
*Assistant Professor of Ophthalmology, Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran, Email: massood.m@gmail.com*

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Herein, we present two cases of early-onset PEX syndrome with different ocular histories.

## Case reports

***Case 1***

A 30-year-old woman was referred to the glaucoma clinic for evaluation of glaucoma and cataract in her right eye. The patient had sustained a penetrating injury with limboscleral laceration and iris prolapse in the right eye when she was 4 years old .The laceration had been repaired and the prolapsed iris excised. At presentation the patient's corrected distance visual acuity (CDVA) was $^{20}/_{60}$ in the right eye. Slit-lamp examination revealed the scar of previous trauma in the inferotemporal limboscleral area, with corresponding traumatic iris defect in the inferotemporal quadrant. There were typical PEX material deposits on the pupillary border and on the anterior capsule of the lens. The crystalline lens was cataractous and mild phacodonesis was noted. Intraocular pressure (IOP) was 15 mmHg on timolol maleate 0.5% twice daily and latanoprost 0.005% daily. Fundus examination was unremarkable with cup-to-disc ratio of 0.4 and a healthy neuroretinal rim. Examination of the left eye was unremarkable with uncorrected distance visual acuity of $^{20}/_{20}$ and no sign of PEX material deposition was seen before and after pupillary dilation.

Phacoemulsification with insertion of a capsular tension ring and in the bag implantation of an acrylic single-piece posterior chamber intraocular lens (PCIOL), and also pupilloplasty with 10-0 prolene sutures were performed on the right eye.

Three months after surgery CDVA in the right eye was $^{20}/_{25}$. On slit-lamp examination PEX material was evident on pupillary border (Figure 1a, 1b), PCIOL was optically clear and posterior capsule was intact. IOP was 22 mmHg on timolol maleate 0.5% and dorzolamide hydrochloride 2% eyedrops.

***Case 2***

The patient was first seen 2 years ago at age 11 at our glaucoma clinic. She had been diagnosed with primary congenital glaucoma in infancy and had undergone angle surgeries in both eyes when she was 6 months old. The right eye had additionally trabeculectomy at age 2 years.

At initial examination her BCVA was $^{20}/_{25}$ in both eyes. Both corneas were enlarged with horizontal diameter of 13 millimeters. Haab's striae were evident in the left eye, inferior and temporal to the corneal center (Figure 1d). In the right eye the filtering bleb was flat. IOP was 36 mm Hg and 15 mm Hg in the right and left eyes, respectively. She was taking timolol maleate 0.5% twice daily, Latanoprost 0.005% daily and Brinzolamide1% twice daily for both eyes. The crystalline lenses were clear and no sign of PEX syndrome was noted on pupillary border or anterior lens capsule. Vertical cup-to-disc ratios were 0.8 and 0.3 in the right and left eyes, respectively. We did a needling bleb revision with mitomycin C on the failed bleb of the right eye. At last follow-up visit, 2 years later, the right eye had IOP of 8 mmHg (with no medications) with an elevated, overhanging filtering bleb superiorly. After pupillary dilation there were flaky white materials on the anterior lens capsule compatible with the diagnosis of PEX syndrome (Figure 1c).The left eye had an IOP of 20 mmHg on timolol maleate 0.5% and latanoprost 0.005% and there was no sign of PEX material deposits on slit-lamp examination.

Although the diagnosis of PEX syndrome in our cases were not confirmed with histopathologic examinations, the deposition of whitish, dandruff-like material on pupillary border and on the anterior lens capsule is quite compatible with the diagnosis of presumed PEX syndrome.

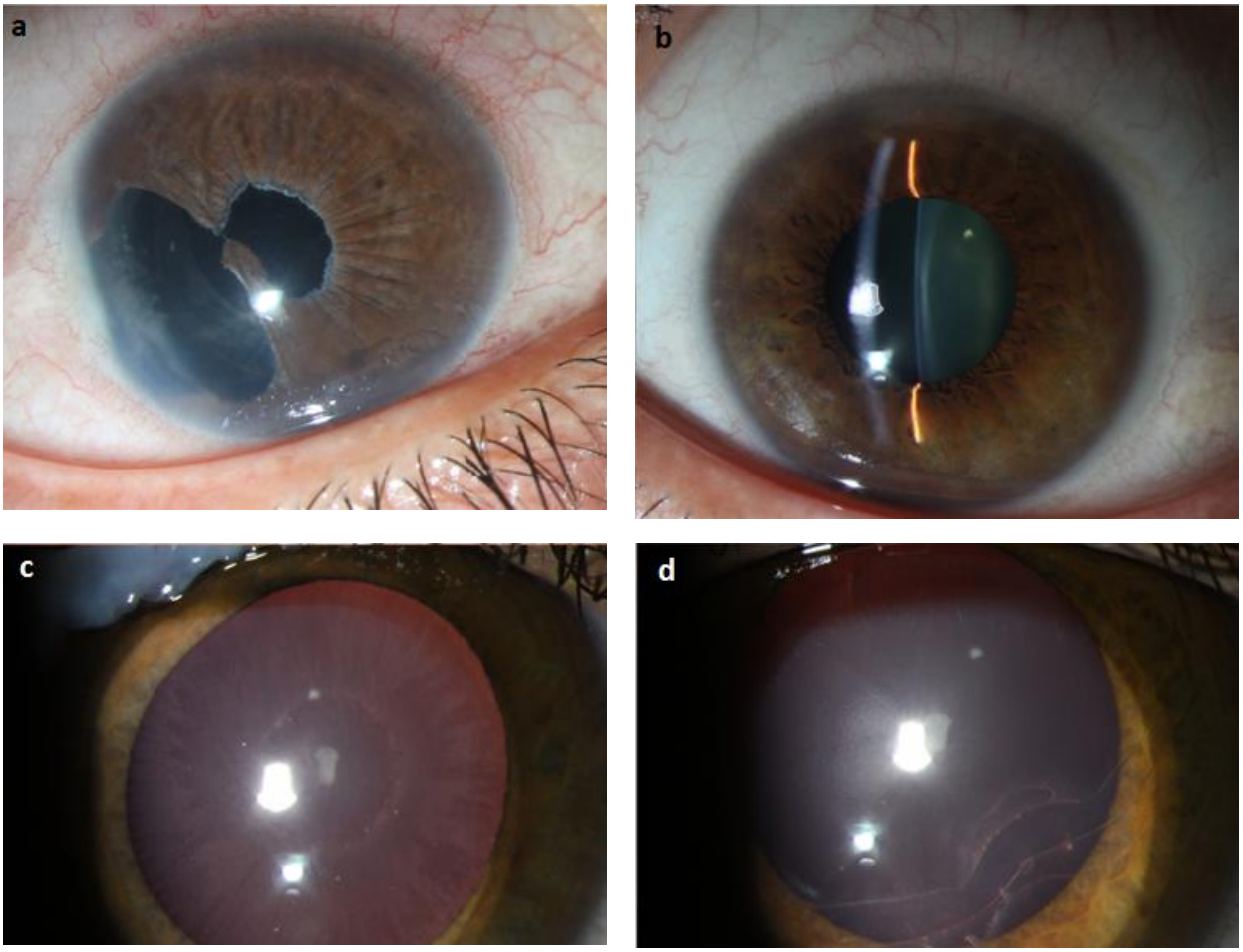


**Figure 1.** Right and left eyes of case 1 (a and b respectively) and case 2 (c and d respectively)

## Discussion

PEX syndrome is an age-related condition and its prevalence increases with age.[1]

Most patients are older than 50 years,[1,2] and there are few case reports of its occurrence before age 40.[2-6] The youngest reported patient with PEX syndrome has been 13 years old.[3] Reported cases were patients with a history of ocular surgery in childhood or early adult age.[4-7] Data regarding the diagnoses, surgeries and ages of young cases with PEX syndrome reported in the literature are summarized in Table 1.

These surgeries usually involved some iris manipulation, mostly a surgical peripheral iridectomy. Similarly, our first patient had sustained iris trauma and the second patient developed PEX syndrome only in the eye that had undergone trabeculectomy (with peripheral iridectomy).

The exact pathogenesis of PEX syndrome remains to be elucidated, but knowledge of the ultrastructural pathology of this process has significantly advanced in recent years.[8-12] These studies have shown that lysyl oxidase-like 1 is a major gene associated with PEX syndrome.

It has been assumed that additional genetic and environmental factors may also have influence on the manifestation of the disease.[13,14]

According to these new developments in understanding the pathogenesis of PEX syndrome, it can be assumed that ocular trauma or surgery in childhood with its associated up- and downregulation of pro-infalmmatory and profibrotic cascades can promote and accelerate the process of PEX syndrome development and accumulation in eyes that are genetically predisposed to PEX syndrome resulting in earlier presentation of this syndrome in young patients.

**Table 1.** Previous reports of pseudoexfoliation syndrome in young patients

| Source (year) | Pts | Initial diagnosis | Intraocular surgery | Age at PEX syndrome detection |
|---|---|---|---|---|
| Yuksel, Schlotzer-Schrehardt, et al (2005) | 1 | [developmental] cataract | ECCE-sulcus PCIOL at age 6 years | 13 |
| Konstas, Ritch, et al (1997) | 1 | Congenital glaucoma | Trabeculectomy in infancy | 17 |
| Kuchle and Naumann (1992) | 2 | Keratoconus | PK at 31,37 and 41 years | 4-6 years after PK |
| Sugar (1976) | 1 | Penetrating trauma, iris prolapse | Repair with iris excision at age 11 months | 26 |
| Horven and Hutchinson (1967) | 2 | JOAG<br>keatoconus | Iridencleisis at age 16<br>PK* at age 24 | 31<br>35 |

Pts: Number of patients, ECCE: Extracapsular cataract extraction, PK: Penetrating keratoplasty, JOAG: Juvenile open-angle glaucoma

## Conclusion

According to a few previous reports of occurrence of PEX syndrome in young patients and surgical histories of our patients, there may be a role for ocular and particularly iris trauma in the development of PEX syndrome in younger patients.

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