

CATARACT SURGERY IN PSEUDOEXFOLIATION SYNDROME

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ABSTRACT: Pseudoexfoliation (PEX) syndrome is an age-related systemic disorder with major ocular manifestations. It is characterized by the production and deposition of fibrillogranular amyloid-like extracellular material within many ocular tissues. Pseudoexfoliation is also a risk factor for glaucoma and also correlated to an increased incidence of cataract formation. Cataract surgery in eyes with pseudoexfoliation is connected with many complications and presents challenges that require careful preoperative planning and intraoperative care to ensure safe surgery and successful postoperative outcome. Zonular weakness and poor pupillary dilation are the two major risk factors for surgical complications and poor visual acuity after surgery. With proper preparation and the use of specialized adjunctive devices, phacoemulsification is the preferred procedure of cataract extraction in this group of patients. Postoperatively, patients require frequent and detailed follow-up to monitor for complications such as intraocular pressure rise, inflammation and intraocular lens dislocation. In conclusion, with appropriate preoperative, intraoperative and postoperative care, the risk of complications can be minimized and favorable outcomes may be achieved in cataract surgery in eyes with pseudoexfoliation syndrome.

KEYWORDS: Pseudoexfoliation syndrome, Cataract surgery, Complications, CTR, Pupillary ring.

INTRODUCTION: Pseudoexfoliation (PEX) syndrome is an age-related systemic disease which have primarily ocular manifestations characterized by the deposition of a whitish-gray pseudoexfoliation fibrillogranular amyloid-like material on the anterior lens capsule (Fig. 1), zonules, ciliary body, pupillary margin, corneal endothelium, anterior vitreous and trabecular meshwork.^{1, 2}

The most important diagnostic sign of PEX is whitish-grey flaky material on the pupillary-border of the iris (Fig. 2) and the anterior surface of lens. The lens usually demonstrates three zones on the anterior lens capsule which consists of a relatively homogeneous central zone and a granular cloudy peripheral zone with a clear zone in between. For detecting these signs a careful clinical examination using dilated slit-lamp bio-microscopy and additionally un dilated gonioscopy are done. Undiagnosed PEX can lead to unexpected problems in management and during surgery.

PEX is a risk factor for open-angle glaucoma, angle-closure glaucoma, lens subluxation, blood- aqueous barrier impairment, and serious intraoperative and postoperative complication and has been correlated with an increased incidence of cataract formation. Jonasson et al. reported a 10% annual increase for both open-angle glaucoma and PEX in persons of 50 years and over in Iceland.³

Although the exact etiology of the condition as well as the exact structure of the material is still unknown. It is presumed that the production of pseudoexfoliation material is associated with abnormal metabolism of glycosaminoglycans and thus abnormalities of the basement membrane in the epithelial cells. Pseudoexfoliation syndrome is prevalent in most parts of the world in varying frequency. Its prevalence increases steadily with age and is rarely seen before the age of 50.

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Recent genetic studies in multiple populations have identified the lysyl oxidase-like1 (LOXL1) gene as one of the major contributor to the risk of developing pseudoexfoliation syndrome and pseudoexfoliation glaucoma.⁴

Preoperative Evaluation: Many studies have stressed that the patients with pseudoexfoliation syndrome have higher rates of complications during and after cataract surgery.^{5,6} The alternations of tissues of the anterior eye segment make cataract surgery potentially challenging and thus surgeons must be aware of numerous intraoperative and postoperative problems in managing the patient with pseudoexfoliation syndrome.

Two pathological manifestations of pseudoexfoliation, zonular weakness and poor pupillary dilation have been identified as the most significant risk factors for surgical complications. A significant zonular instability can cause phacodonesis,⁷ spontaneous subluxation of the lens⁸ (Fig.2) and angle-closure glaucoma due to pupillary and ciliary block.⁹ Preoperative reduced anterior chamber depth indicates zonular instability and should therefore alert the surgeon to the occurrences of intraocular complications related to zonular dialysis. Patients should be evaluated for presence of glaucoma and a triple procedure combining cataract surgery and trabeculectomy may be planned.

The reduced pupillary dilatation and zonular fragility also leads to complications such as posterior capsular rent, vitreous loss and retained cortical material. Pseudoexfoliation syndrome influences the development of synechiae between the iris pigment epithelium and the anterior lens capsule due to blood-aqueous barrier breakdown. Posterior synechialysis or lysis of more peripheral iridocapsular adhesions and pupillary enlargement are often necessary.¹⁰

Steps of Surgery: Phacoemulsification is the preferred surgical technique but its use should be directed by the experience of the surgeon. Many studies showed a lower complication rate with phacoemulsification than extracapsular cataract extraction in eyes with pseudoexfoliation.^{5,11,12}

Inadequate pupillary dilation and zonular laxity are the important factors to be dealt with while performing the surgery.

Small Pupil: In small pupils, various methods are employed to produce adequate dilatation. Posterior synechiae warrants synechialysis. Highly cohesive ophthalmic viscosurgical devices [viscomydriasis], sphincterectomy, mechanical iris dilators (Fig. 3) such as pupillary hooks and pupillary rings (Fig. 4) can be used to dilate pupil. Iris hooks can also serve to support anterior capsulorhexis if zonular weakness is significant. Residual sphincter damage can permanently dilate the pupil.

Capsulorhexis: Capsulorhexis should be performed with a sharp instrument. Presence of striae during anterior capsular puncture is an indication of zonular weakness. Usage of iris hooks for counter traction on anterior capsule may be necessary in some cases during capsulorhexis.

Capsulorhexis size an important factor and it should be performed within the limits of the outer pseudoexfoliation ring. Too small a diameter will put further stress to loose zonules during the operation while too large a diameter may engage the zonular attachments.

Capsular tension Ring: As already emphasized, the complications during phacoemulsification surgery are mostly related to zonular weakness and therefore it requires particular attention.

Depending on zonular weakness or dialysis a capsular tension ring (CTR) may be inserted prior to phacoemulsification or after phacoemulsification of nucleus and prior to cortical irrigation-aspiration. In some difficult cases, Cionni modified-CTR (mCTR) or capsular tension segments (CTS) may be inserted either alone or in combination. When the degree of zonular weakness is mild, a CTR (Fig.5) may be enough to provide support intraoperative manoeuvres and to stabilize an IOL in the capsular bag.

If obvious phacodonesis is present, then it is advisable to insert a CTR on completing the capsulorhexis to stabilize the capsular bag for phacoemulsification and cortical removal. For more advanced zonular instability the mCTR or CTS both of which can be sutured to the sclera for improved fixation may be considered. Unlike the CTR (Fig.6), the CTS do not require a dialing technique during insertion and therefore produces much less stress on the zonules. With respect to all these facts, the entire surgical procedures must be conducted with precise care.^{13,14,15}

During the operation it is also paramount to avoid overinflating the anterior chamber with saline solution or viscoelastics to avoid undue stress on the zonules. Hydro dissection should be carried out judiciously with minimal stress on the zonules.

Phacoemulsification: With regards to the phacoemulsification technique, it is recommended that each surgeon applies his or her technique of preference. For denser nucleus, horizontal chop technique is the preferred whilst for softer nucleus it is the anterior phaco technique. In order to avoid zonular stress adjust machine settings in the medium range (vacuum 250-300 mm mercury, aspiration flow rate 20-30 cc/min, US power 50-60% continuous). Cortex removal is a critical step and it should therefore be done with tangential stripping rather than centripetal movements thus aiming to avoid transmitting pressure to zonules and posterior capsule. In cases with inserted CTR (Fig 5), cortical remnants may get entrapped which presents an additional obstacle for the cortical removal.

Intraocular lens Implantation: The selection of intraocular lens (IOL) is also important in the eyes with pseudoexfoliation syndrome. Heparin surface modified posterior chamber IOLs have been found to be associated with fewer postoperative fibrinoid reactions, lesser pigment and cellular deposits on the lenses and lower incidence of the posterior synechiae formation than other forms of IOLs.¹⁰ In the case of severe zonular instability and dialysis or loss of vitreous an alternative option are scleral, ciliary sulcus or iris-fixated IOL.

After surgery, the pseudoexfoliation patients must preferably undergo thorough and frequent examinations for the early detection of an increase of IOP, inflammation and late onset IOL dislocation. Administering acetazolamide immediately after the surgery and using topical glaucoma mediators in the postoperative period can be effective in the control the rise of IOP. In addition, it is necessary to use topical steroids and NSAIDs to control inflammation which tends to be higher in these patients.^{15,16}

The process of pseudoexfoliation continues even after the surgery. So these patients must be monitored for the possible development of glaucoma, capsular phimosis syndrome or decentration of IOL. There is also the possible risk of future IOL-CTR capsule complex dislocation which may require surgical intervention.

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COMPLICATIONS: Pseudoexfoliation is associated with increased occurrence of complications, both during and after cataract surgery.¹⁷ experienced surgeons produce far less complications compared to junior or resident surgeons.

A) Intraoperative Complications: Inadequate pupillary dilation and zonular laxity are the important factors that lead to complications. Damage to iris sphincter, residual lens material, hyphaema and iridodialysis occur frequently. Zonular laxity produces problems such as zonular dialysis, posterior capsular rupture and vitreous loss.¹⁷ Usage of capsular supporting devices¹⁸ as well as iris hooks can help the surgeon reduce these complications.

B) Postoperative Complications: Severe anterior chamber reaction is seen frequently. It should be differentiated from endophthalmitis. Increased intraocular pressure and corneal edema are also encountered. These should be identified and treated promptly. Hyphaema from iris is usually seen in the immediate post-operative period. Pigment dispersion and residual lens material may occur. These patients are more prone for the development of glaucoma, posterior capsular opacification, IOL tilt or dislocation.¹⁹ Therefore it is advisable to do long term follow up of post cataract surgery pseudoexfoliation patients.

CONCLUSION: Pseudoexfoliation syndrome presents many challenges that need careful preoperative planning and intraoperative care that will ensure safe surgery and a successful postoperative outcome. With adequate preparation and use of specialized adjunctive devices, phacoemulsification is the preferred method of cataract extraction in eyes with pseudoexfoliation syndrome. In addition, proper follow-up of patients after surgery is needed to evaluate endothelial cell function, intraocular pressure rise, inflammation and intraocular lens dislocation. Conclusively, the risks associated with cataract surgery in eyes with pseudoexfoliation can be minimized with appropriate preoperative, intraoperative and postoperative care.

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Fig. 1: Image pseudoexfoliative material over the lens surface.

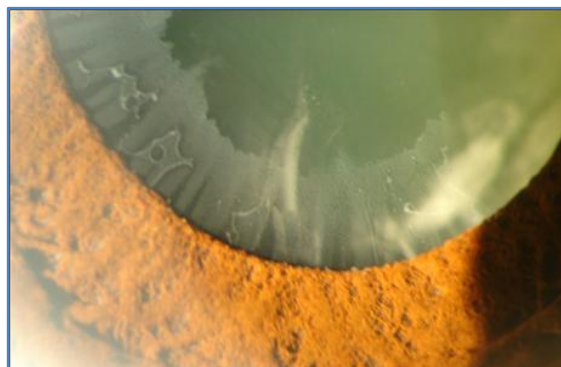


Fig. 1

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Fig. 2: Image shows pseudoexfoliative material on pupillary margin and inferiorly subluxated lens.

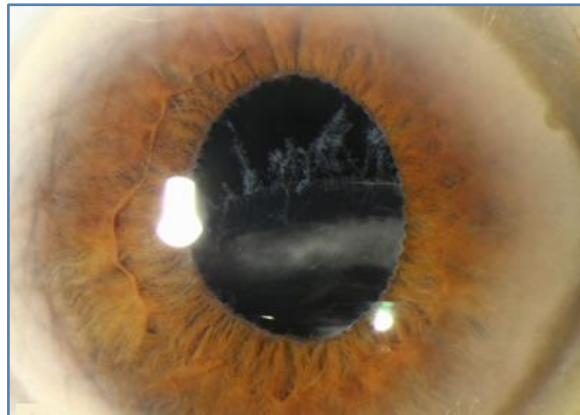


Fig. 2

Fig. 3: Iris hooks

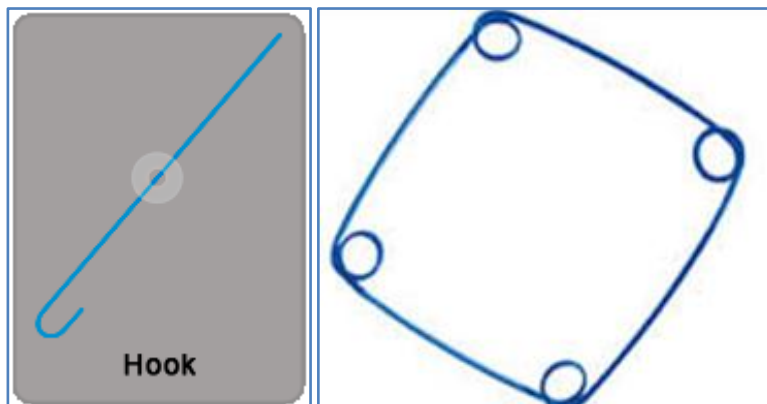


Fig. 3

Fig. 4: Malyugin Ring being used in surgery to keep the pupil dilated.

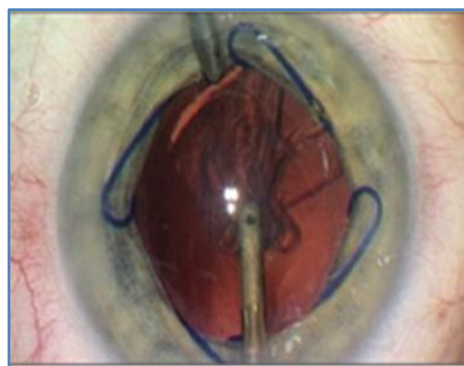


Fig. 4

Fig. 5: Capsular tension ring

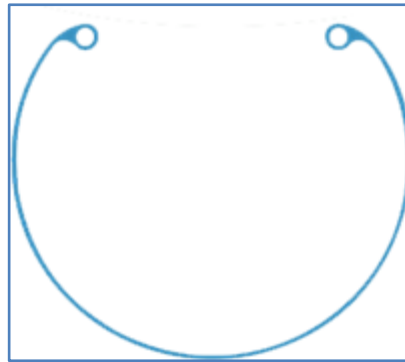


Fig. 5

Fig. 6: Diagrammatic representation of usage of capsular tension ring.

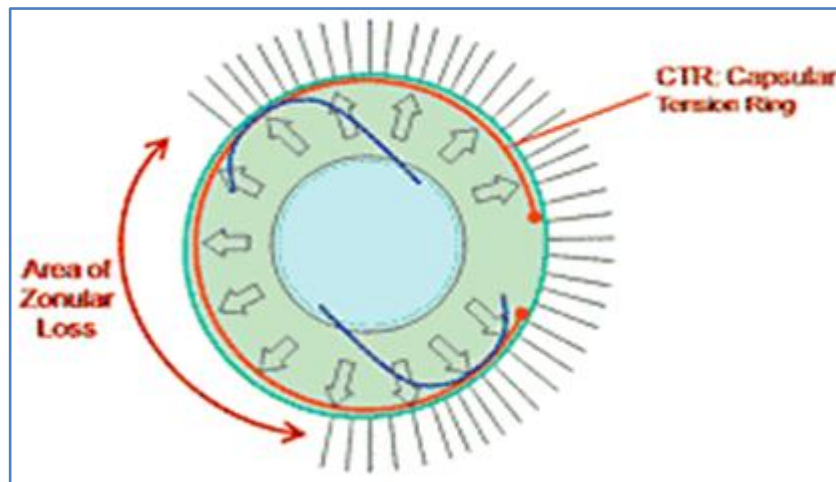


Fig. 6

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