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Case Report

# Dislocated Posterior Chamber Intraocular Lens (PCIOL) in Patients with Retinitis Pigmentosa (RP)

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**Abstract**

Subluxation or dislocation of PCIOL is one of the complications of cataract operation in RP patients. This paper reports the presentation of PCIOL dislocation and subluxation and the management and outcome in 3 eyes of 2 RP patients. Two medical records of patients with RP who developed dislocated or subluxated PCIOL and subsequently underwent explantation of the dropped IOL were evaluated. Two patients had bilateral eye cataract operation done and had PCIOL implanted. Patient 1 developed left eye subluxated PCIOL inferiorly after 2 years of the cataract operation and right eye dislocated PCIOL anteriorly 4 years after cataract operation. Patient 2 develop right eye subluxated PCIOL inferiorly after 12 years of the cataract operation. Patient 1 with right eye dislocated PCIOL underwent intraocular lens (IOL) explantation and was left aphakic as her visual prognosis was poor due to advanced RP. The left IOL remained within the visual axis despite subluxation and no intervention has been done. Patient 2 with right eye subluxated PCIOL underwent IOL explantation and anterior chamber intraocular lens (ACIOL) implantation. ACIOL remained stable and visual acuity improved post-operation. Both the operations were uneventful. Post-operatively, there was no elevated intraocular pressure and no prolonged ocular inflammation, which required prolonged anti-inflammatory and no retinal detachment was seen. Both patient and surgeon should be aware of potential PCIOL subluxation or dislocation in RP. The presentation may be as late as more than a decade after the cataract operation.

**Keywords:** Artificial lens implant migration, aphakia, cataract extraction, lens subluxation, pseudophakia, retinitis pigmentosa

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## Introduction

Retinitis pigmentosa (RP) is a genetically heterogeneous group of retinal dystrophies characterized by progressive dysfunction of predominantly rod photoreceptors, with subsequent degenerations of cones and retinal pigment epithelium. RP is classically characterized by retinal pigmentary bony spicules changes, arteriolar attenuation and waxy disc pallor; while patient with RP often presented with night blindness and progressive peripheral visual loss. A common sign of RP is posterior subcapsular cataract, which develops at a relatively earlier age, and may cause significant visual impairment (1). Despite the potentially poor visual prognosis in RP, cataract surgery is often required and patient may have major improvement in visual acuity after cataract surgery provided the macula retains some visual function (2).

IOL dislocation or subluxation is a devastating complication that may follow cataract operation. The incidence of such complication ranges from 0.2% to 3.0% (3,4). Previous case reports have documented dislocation or subluxation of in-the-bag foldable

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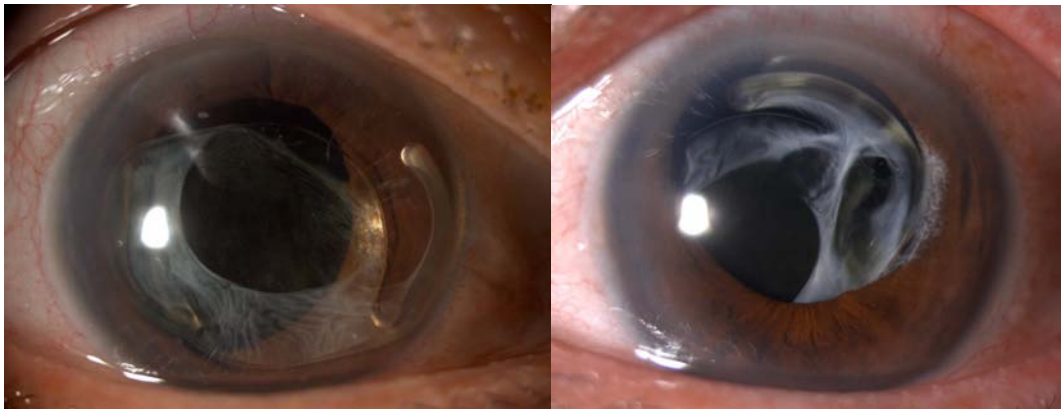

(a) (b)

**Figure** 1: a) Anterior segment photograph of the right eye show inganterior dislocation of in-the-bag PCIOL; b) Anterior segment photograph of the left eye showing inferior subluxation of the PCIOL

intraocular lens (IOL) following uneventful cataract operation in RP patients (5,6). Various mechanism have been postulated, and this may be associated with post-operative capsular shrinkage and zonular weakness or dehiscence (6-8). This may lead to poor visual outcome of cataract operation in RP patients.

Here we report retrospectively 3 eyes of 2 RP patients who developed dislocated IOL following cataract operation.

## Case Report

*Patient 1*

A 60-year-old Chinese lady was referred to vitreo-retinal service for right eye dislocated PCIOL into the anterior chamber and left eye subluxated PCIOL. She was diagnosed with retinitis pigmentosa in a private center when she was 52 years old and subsequently had bilateral eye cataract extraction done with single piece acrylic IOL implanted in the bag in 2007. The operation was otherwise uneventful.

In 2009, the patient was referred to Universiti Kebangsaan Malaysia Medical Center (UKMMC) for low vision aids and was noted to have left eye PCIOL inferior subluxation. The patient's RP was already in an advanced stage upon presentation as there were extensive bony spicules bilaterally with pale optic disc. Visual acuity was hand movement (HM) bilaterally. No surgical intervention was planned for her left eye as the IOL was still within the visual axis and appeared stable.

In 2011, 4 years after the cataract operation, the patient presented with right eye sudden onset of blurred vision. On examination, the best-corrected visual acuity (BCVA) was light perception (PL) in the right eye, with no relative afferent pupillary defect (RAPD). In the right eye, there was anterior dislocation of the PCIOL into the anterior chamber (Fig. 1a). Cornea was clear otherwise and intraocular pressure was normal. In the left eye, the subluxated PCIOL remained stable, with the superior haptic tilted anteriorly while the other haptic remain in the bag (Fig. 1b).

The patient was offered right eye IOL explantation with repositioning of IOL, however, she was not keen due to personal reasons. The operation was delayed till 2015, when patient eventually had right eye IOL explantation with anterior vitrectomy done. There was no lens implantation done, as her visual prognosis was poor due to her advanced RP. As for the left eye, the subluxated IOL remain stable with no further subluxation or dislocation, and therefore, no operation was planned. Post-operatively, the right eye vision remained poor at hand motions, even with aphakic addition.

*Patient 2*

A 47-years-old Malay lady initially presented to UKMMC in 2002 with bilateral eye progressive painless visual loss. She was treated as bilateral eye uveitis and subsequently had left eye cataract extraction done with PCIOL (single piece acrylic IOL) implantation in 2003. There were no features of retinitis pigmentosa noted at the time.

The patient then defaulted her follow-up from 2008 till 2015, when she presented again with sudden onset of blurring of vision and monocular diplopia in the left eye, 12 years after the cataract operation. On examination there was left eye subluxation of PCIOL inferiorly out of the visual axis with superior zonulolysis.

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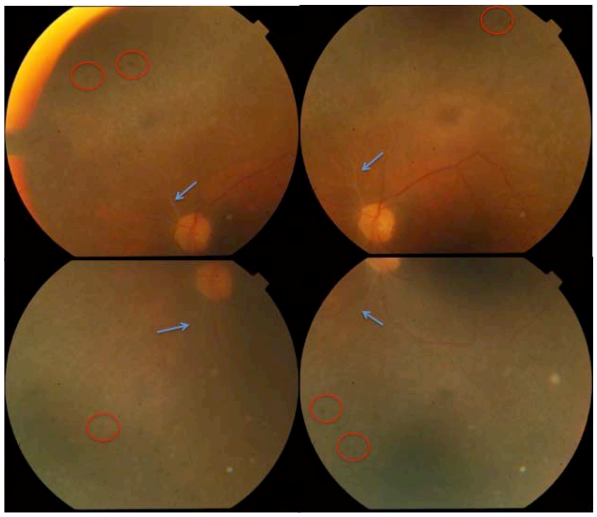

**Figure 2**: Colour fundus photograph showing changes suggestive of RP: bony spicules (red circle), antenuated vessels (blue arrow) and disc pallor

Visual acuity of the eye was counting finger (CF) and there was no RAPD. Intraocular pressure was normal. Upon fundus examination, bony spicules were noted in the retina periphery with palish optic discs bilaterally. Therefore, she was diagnosed as retinitis pigmentosa, which was evident by the clinical findings (Fig. 2).

The patient subsequently underwent left eye IOL explantation with anterior chamber IOL (ACIOL) implantation. Post-operatively, left eye vision improved to 6/36 and the ACIOL remained stable.

Patient also had right eye cataract operation done in 2010 in private center and the IOL has so far remained in the bag.

**Discussion**

RP is one of the main predisposing factors in dislocation or subluxation of PCIOL after cataract operation, apart from pseudoexfoliation (9). It can happen both as an early or late complication. Previous case studies have reported the complication presenting as early as weeks to many years after the operation (5,6).

Various mechanisms have been proposed to be associated with IOL dislocation in RP such as zonular weakness and anterior capsular contraction (6,7). Prior case reports have noted zonular loss and laxity on ultrasound biomicroscopic examination preoperatively, and subsequently presented with IOL dislocation later postoperatively (6). Several reports also noted zonular weakness or dehiscence intraoperatively and postoperatively (5). The phacoemulsification process may have further aggravated the zonular weakness. In this small case series, there was no documented phacodonesis prior to the operation. Hayashi et al. had demonstrated that there is significant reduction in the mean area of anterior capsular opening after 1 month of cataract operation in RP patients compared to control group. Some of the patients had decrease opening area of less than 10mm2, which required anterior capsulotomy (7). The cause of this unusual phenomenon in RP patients is unknown.

Knowing that IOL dislocation can happen in RP patients, various precautions may need to be taken during cataract operation. Hayashi et al. suggested a larger size of capsulorhexis during cataract operation might decrease the risk of anterior capsular contraction and subsequent IOL dislocation or subluxation (7). Capsular tension rings (CTR), which are indicated in cases where there is zonular dehiscence or rupture intraoperatively, may be used to reduce post-operative capsular shrinkage (8). However, there have been case reports of IOL dislocation despite CTR implanted in RP patients (10). CTR implantation may also make explantation of the lens more challenging.

Stress and damage to the zonules can also be minimized by adopting ideal phacoemulsification techniques. For instance, a phaco-chop technique may be used to minimize stress on the zonules, thus preserving the integrity of the zonules (8). Post-operatively, capsular contraction should be documented and followed up. For this, radial Nd:YAG laser anterior capsulotomy may be performed if warranted (7).

Vitreoretinal surgery involving explantation of the IOL is needed when there is subluxation or dislocation of IOL resulting in visual loss in the eye or when the dislocated IOL may potentially cause retinal or corneal pathology. Subluxated IOLs which remain in the visual axis should be observed, as demonstrated in the left eye of patient 1. Other eyes may need removal of the IOL with or without implantation or repositioning of the IOL. Generally the indication of surgical intervention includes reduction in visual acuity, diplopia, glaucoma, risk of corneal decompensation (in the case of anterior lens dislocation) or retinal detachment (8).

The most common surgical corrections for subluxated or dislocated PCIOL are IOL repositioning with ACIOL and posterior scleral fixated IOL (PSFIOL) (11). In patient 2, she had right eye ACIOL implantation. Various studies had compare the outcome of the both technique, both with its own pros and cons (11,12). ACIOL are associated with

complication like post-operative corneal decompensation and raised intra-ocular pressure, due to the close contact of the lens with the corneal and angle structures (11,12). Both of this complication can cause worsening of vision. Different techniques had been described for PSFIOL, including continuous-loop fixation, four points fixation, transcleral fixation through sclerotomy, scleral tunnel technique, etc (13). The complication includes intraocular haemorrhage, retinal detachment, suture erosion and subsequent IOL decenteration (11,12). Some studies have reported higher complications in PSFIOL compare to ACIOL (12). Other studies however, suggested there was no significant difference between both techniques (11). PSFIOL implantation is more challenging to perform and requires an experienced surgeon while ACIOL implantation is relative more easy and safer to perform. It is suggested that a sufficient amount of viscoelastic during intra-operative manipulation to protect the endothelium may help to reduce post-operative complication in ACIOL implantation.(12) Many studies have demonstrated no significant difference in mean visual acuity between the two surgical techniques (11,12). Therefore, in view of the similar outcome, patient 2 underwent ACIOL implantation.

RP is a progressive disease. Various studies have suggested the rates of decline are associated with mode of inheritance, possible modifier genes, environmental and dietary factors and stage of disease (1). It is reported that autosomal dominant RP has the best prognosis while x-linked recessive had the worst prognosis. Autosomal recessive or sporadic cases were intermediate in severity (1). In a study involving 999 patients with RP age 45 years and older, 25% of them had a visual acuity of 20/200 or worse in both eyes while 0.5% of them had no light perception in both eyes (14). This indicates that RP patients rarely will lose all vision in both eyes, however, majority of them will required low vision rehabilitation. Low vision rehabilitation includes magnification, optical aids such as telescope, centric viewing and reading techniques. In a study looking into visual rehabilitation among patients with advanced stage of glaucoma, optic atrophy, myopia and RP, there was significant improvement in RP group, though the improvement was the least prominent (15). In patient 1, the patient was in an advanced stage of RP as evidenced by the extensive bony spicules reaching the macula with thin vessels and pale optic disc, therefore, patient had poor visual prognosis and IOL explantation was done without secondary IOL implantation. Patient subsequently was referred for low vision aids.

## Conclusion

In conclusion, RP predisposes to PCIOL subluxation or dislocation post cataract operation. Patients require surgical management in the form of vitrectomy surgery, in most cases with removal of dislocated IOL and visual rehabilitation with secondary IOL or visual aids depending on the severity of RP and the expected visual outcome. Counselling on this complication in RP patients should be given prior to the initial cataract surgery to alert patients on the symptoms and the options for recovery of vision should dislocation occur.

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