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REVIEW/UPDATE

Cataract surgery in patients with retinitis pigmentosa: systematic review

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Retinitis pigmentosa (RP) is an inherited bilateral retinal degenerative disease with an incidence of 1 in 4000 people. RP affects more than 1 million individuals worldwide. Although night blindness and restricted visual field are the most typical symptoms of these individuals, generalized vision loss due to cataracts can be expected in the latter stages of the disease. It has been demonstrated that posterior subcapsular cataract is the most prevalent cataract in younger individuals with RP, as opposed to age-related cataracts. Although most ophthalmologists may have a negative view of cataract surgery in patients with RP, it appears that it can play an important role in the visual restoration of patients with RP. However, there are concerns about performing cataract surgery for patients with RP. Herein, a systematic review was conducted according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses on databases of MEDLINE and Scopus.

J Cataract Refract Surg 2023; 49:312–320 Copyright © 2022 The Author(s). Published by Wolters Kluwer Health, Inc. on behalf of ASCRS and ESCRS

Retinitis pigmentosa (RP) is a hereditary degenerative disease that affects the retinal pigment epithelium and photoreceptors, causing progressive night blindness and visual field narrowing throughout the early decades of life.[1,2] The global prevalence of RP has been estimated to be 1 in 4000, affecting more than 1 million individuals.[1] Multiple inheritance patterns of RP influence the onset age and severity of symptoms. RP is often bilateral and characterized as nonsyndromic or simple (not affecting other organs or tissues), syndromic (affecting other neurosensory systems such as hearing), or systemic (affecting multiple tissues).[1,3] Central vision will not be lost until the end stages, which include direct macular involvement. Other causes of central vision loss in RP include cystoid macular edema (CME) and cataract.[1]

Cataract formation is a common problem in RP that occurs at a younger age than the age-related types, especially in the X-linked form of RP.[4,5] Although management of cataracts in patients with RP can considerably aid in the restoration of vision, it may be accompanied by certain complications that may worsen the visual condition of these patients. This study aims to review the different aspects of cataract surgery in patients with RP considering preoperative evaluations, intraoperative precautions, and postoperative complications.

METHODS

The systematic index review was conducted according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses, 2009. Two authors searched central databases, including MEDLINE and Scopus, for the following keywords: "cataract surgery," "phacoemulsification," and "retinitis pigmentosa." The search results were confined to human studies published since 2000; the first literature search was performed on November 1, 2020.

After the initial search, the first screening of articles was performed by a rapid review of article topics. Selected papers underwent a second screening by reviewing their abstracts. The screening process was performed by 2 reviewers independently, and any discrepancies in their results were resolved by discussion between the reviewers. The reviewers were not masked to publications and the authors' names. The full texts of relevant articles were provided for evaluating their eligibility and data extraction. References were also obtained from citations found in the original search. The last search was conducted on July 14, 2022, to encompass the latest related published articles. The initial search found 172 papers, of which 42 were duplicates. After screening topics and abstracts, the full text of 42 articles was extracted and reviewed for data collection. Most papers were case reports and case series, whereas only a few comparative studies were yielded. Therefore, to cover the issue comprehensively, additional references regarding eyes without RP were used to extrapolate information related to patients with RP.

Cataract Epidemiology Among Patients With RP

One of the most common conditions complicating RP is the development of cataracts in which posterior subcapsular cataract

Submitted: May 23, 2022 | Final revision submitted: November 9, 2022 | Accepted: November 13, 2022

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0886-3350/$ - see frontmatter
https://doi.org/10.1097/j.jcrs.0000000000001101

(PSC) is the most common type.[6,7] About 2% of people aged 52 to 85 years may demonstrate PSC, whereas 50% of patients with RP develop PSC at younger ages than age-related cataracts.[8] The odds of PSC formation is 41% to 90% by age 40 years and can increase to 80% by age 60 years. The mean age of cataract surgery in patients with RP is 47 to 58 years compared with 72 to 74 years for age-related cataracts in the general population.[4,5,8] However, the mean age of surgery was 62.6 years in 1 study, which can be related to the sensitivity threshold in deciding to undergo surgery.[9]

Underlying Pathophysiology

The definite pathophysiology of cataract formation in patients with RP is unknown.[10] In the normal eye, the lens epithelium is a physiologic barrier regulating the transfusion of different nutrients and ions between aqueous humor and lens fibers.[11] Malfunction of this mechanism can cause intralenticular fluid accumulation and subsequent cataract development.[12] On the slitlamp examination, some patients with RP and PSC show small dots and channel-like structures on the anterior capsule.[10] Of interest, a study on the anterior lens epithelium by electron microscopy has revealed numerous holes as large as 50 μm. It has been speculated that these holes may act as a passageway to conduct leaked fluids into the crystalline lens. Lens fiber cells can transfer this flow to the posterior pole, where alterations in function and structure may lead to cataract formation.[10] In addition to abnormal lens epithelium, several pathologies in the lens fibers of patients with RP with cataracts are discussed. Severe disarrangement of the lens fibers leading to distorted membrane features and cytoplasmic deposits were introduced by Eshaghian et al. in 1980.[13]

Many studies addressed the proinflammatory status of the intraocular microenvironment in patients with RP by evaluating serum, aqueous humor, or vitreous samples.[14–16] These studies reported increased levels of interleukin (IL)-2, IL-6, IL-10, interferon γ, matrix metalloproteinase, and inflammatory chemokines.[14–16] It is hypothesized that these proinflammatory cascades may be produced from degenerated retinal tissue and diffused through the disrupted blood–retinal barrier.[17] Diffusion of these cytokines and chemokines into the lens fibers may interfere with the homeostatic functions of lens epithelial cells, leading to lens metabolic disorders and the migration of fiber cells.[18]

Trends of the Ophthalmologists

It seems that most ophthalmologists may not be entirely enthusiastic about performing cataract surgery in patients with RP. In one of the studies, a 10-item questionnaire was sent to Japanese ophthalmologists.[19] Among 197 responders, only 18.1% recommended cataract surgery in RP, focusing on the severity of cataracts, the patient's desire, and the degree of visual acuity (VA). Notably, 56.6% of responders stated that cataract surgery in these patients might be harmful.[19] Some authors discussed the risk/benefit ratio of cataract surgery, especially for the risk of increased photophobia and removing physiologic photoprotection attributed to the presence of the crystalline lens.[20] On the other hand, a wait attitude regarding cataract surgery is more welcome.[20]

Effects of Cataract Surgery on Retinal Anatomy and Physiology

Garcia-Martin et al. evaluated anatomic and neurophysiologic tests of 35 patients with RP diagnosis 1 month before and 1 month after uneventful cataract phacoemulsification. These tests included visual evoked potential (VEP), pattern electroretinogram, as well as retinal nerve fiber layer (RNFL) and macular thickness measurements.[21] They also compared image repeatability preoperatively and postoperatively using Cirrus and Spectralis optical coherence tomography (OCT) in this observational prospective longitudinal study. They found no significant differences in the pattern electroretinogram values (amplitude and the latency of P-50 and N-95) and VEP latency after cataract surgery, whereas the VEP amplitude was increased postoperatively (P = .03). In addition, OCT scans showed a postoperative increase in RNFL thickness and central macular thickness provided by the Cirrus OCT and improved repeatability of OCT measurements (P < .001) after cataract removal in patients with RP. By contrast, macular thicknesses measured by Spectralis OCT preoperatively did not differ significantly from that postoperatively. The authors suggested that when using Cirrus OCT, ophthalmologists must consider the variability in measurements associated with the cataract surgery as a probable cause of changed macular thickness postoperatively, whereas using the Spectralis OCT the increased macular thickness detected postoperatively should be attributable to potential retinal inflammation or postsurgical edema, rather than the device variability (perhaps because of the TruTrack technology used by Spectralis OCT). However, the authors did not provide further evidence to support the idea of a correlation between RP progression after cataract surgery and increased macular thickness. Furthermore, they concluded that both Cirrus and Spectralis OCTs have better repeatability after cataract surgery; therefore, the values measured with OCT in patients with RP in the presence of cataracts should be interpreted cautiously. The changes in macular or RNFL thicknesses might be attributable to the test's variability associated with the presence of cataracts and not caused by an actual change in the RP pathology.[21]

An electrophysiological study was conducted by Wang et al. evaluating 34 eyes of 21 patients; no significant change was seen in the electroretinogram of patients with RP after cataract removal, whereas the visual field improved in 17% of studied eyes (6/34).[22]

Preoperative Evaluations

Phacodonesis The prevalence of zonular laxity is significantly higher in patients with RP.[4] A relatively small percentage of patients (3.75%) may show signs of phacodonesis during the preoperative examination, which can allow the surgeon the opportunity to consider the use of techniques that minimize stress on the zonular fibers during cataract surgery.[4] It has been hypothesized that several factors may increase the rate of zonular weakness in patients with RP. In addition, degeneration of photoreceptors is associated with an increased production of inflammatory cytokines, which can potentially damage zonular fibers.[16,23] Similar to the pathophysiology of developing PSC in RP, anteriorly spreading of toxic materials from the degenerated retina can lead to zonular injury.[24] Also, the role of capsular phimosis in developing zonular insufficiency is remarkable. However, capsular phimosis can be considered one of the consequences of zonular weakness.[25,26]

Cystoid Macular Edema CME is another challenge in managing patients with RP. The prevalence of CME in patients with RP is about 20% to 50%, with more recent studies showing higher rates of CME found on OCT than those previously reported with fluorescein angiography (FA).[27–30] It should be noted that the most sensitive diagnostic test to detect CME in patients with RP is OCT; cystoid spaces in these patients may demonstrate no evidence of leakage or pooling on FA.[31]

De Rojas et al. evaluated the outcomes of cataract surgery on 19 eyes with RP. They reported that of 6 eyes with OCT-detected CME during the study, 4 eyes had preoperative CME. In comparison, the other 2 eyes were complicated by postoperative CME or pseudophakic CME (PCME).[32] Although the authors reported no decreased corrected distance VA (CDVA) in any of the studied eyes, the existence of preoperative CME may interfere with the restoration of VA and expectations of the patients and surgeons from the surgery. Hence, evaluating for CME by OCT imaging before the surgery should be considered by physicians who plan to perform cataract surgery in patients with RP.

Visual Prognosis After Cataract Surgery Visual outcomes can be challenging to predict when both lenticular and retinal pathologies exist. Of note, surgery in the RP population poses risks because of complications of postoperative posterior capsular opacification, aggressive anterior capsular contraction, zonular weakness, intraoperative macular phototoxicity, and CME.[1]

Various studies have shown the beneficial effects of cataract extraction on patients with RP (Table 1).[4,5,9,21,22,32–42]

Evaluating a series of 142 eyes of 89 patients with RP undergoing cataract surgery, Jackson et al. demonstrated that vision has improved in these patients after the surgery (logMAR acuity: 1.05 ± 0.38 to 0.63 ± 0.49).[5] They reported that 77% of the eyes improved, 20.5% unchanged, and 2.5% deteriorated in the CDVA; 96.6% increased subjective vision overall. Patients who did not have improved vision had worse preoperative vision than others. It should be noted that this study was conducted between 1985 and 1997. With the advent of phacoemulsification, the percentage of vision improvement may be even higher.

Chan et al. conducted a survival analysis to investigate the duration of visual improvement after cataract extraction in 67 eyes of 42 patients with RP with <10 degrees (64.2% of patients) and ≥10 degrees (35.8% of patients) of visual field.[34] They concluded that most patients had significant improvement of CDVA preoperatively (1.27 ± 0.42 preoperatively to 0.92 ± 0.49 and 0.97 ± 0.53 at 3 months and 1 year postoperatively, $P < .001$), and the mean duration of visual improvement after cataract surgery was 8.10 ± 0.83 years. There was no significant difference in the course of visual improvement between the 2 groups ($P = .345$). The authors showed that these patients could achieve visual improvement over a significant amount of time after cataract surgery, independent of disease severity.[34]

Yoshida et al., in a retrospective study with 56 eyes of 40 patients, investigated patients with RP who underwent cataract surgery over a 5-year follow-up and reviewed factors that affected the postoperative VA.[9] Based on automated static perimetry (Humphrey Field Analyzer) 10-2 and the mean deviation (MD) value, patients were divided into 2 groups: advanced RP with MD less than −15 and less advanced RP with MD greater than −15. Macular OCT was performed for all patients to evaluate the complications such as CME and epiretinal membrane (ERM). Additionally, ellipsoid zone (EZ) disruption at the fovea was assessed and graded; grade 1, the EZ was not visible; grade 2, abnormal EZ; and grade 3, normal EZ. They investigated the relationship between the final postoperative CDVA and all these factors. After data analysis, they reported that the final CDVA was significantly better in less advanced RP with MD greater than −15 and in patients with EZ grade 3 ($P \leq .005$). They concluded that the preoperative MD value and EZ grade might be critical prognostic factors in patients with RP.[9]

Several studies have demonstrated that retinal microstructure status could assist in predicting visual outcomes of cataract surgery in patients with RP.[9,37,39] The width of the EZ (previously called the inner segment and outer segment line), measured by macular OCT, has been demonstrated to be a reliable marker of RP severity and a good predictor of visual outcome after cataract.[9,37,39] Intact external limiting membrane and relatively normal central macular thickness have also been reported to be good prognostic factors.[37]

Noninvasive estimation of retinal blood flow was also postulated for differentiating between the retinal disease and the cataract origin of VA loss in patients with RP before cataract surgery.[43] In an OCT angiography (OCTA) study, the authors reported significantly lower superficial and deep flow density in the parafoveal region compared with healthy controls. In a multivariate analysis assessing predictive parameters of OCT and OCTA on VA, deep parafoveal flow density and the superficial foveal avascular zone were significantly correlated with VA.[43]

Jackson et al. reported that patients with unchanged VA after cataract surgery had worsened preoperative CDVA than the others.[5] Dikopf et al. also demonstrated that VA was significantly improved postoperatively in patients with RP, with a preoperative vision of 20/200 or better.[4] Conversely, patients with a preoperative VA of 20/400 or worse had less objective improvements, maybe because of macular involvement.[4]

However, in the study by Davies et al. of a group of RP and RP-like patients (Usher syndrome, Refsum disease, and Leber congenital amaurosis), only 1 eye of 30 (3.3%) had VA that remained unchanged after cataract surgery (all other eyes had improvement in vision).[35] Visual acuities improved significantly in both subgroups of those with preoperative vision 20/200 or worse and those with preoperative vision 20/150 or better ($P = .001$ and $P = .0002$, respectively), rather than VA improvement being limited to patients with better preoperative vision. They suggest that the selection of cases for cataract surgery can effectively be made based on the patient's report of symptoms, specifically decreased central vision or increased glare.

Patients with RP also suffer from glare in addition to low VA, which contributes further to decrease in visual function. These patients' primary sources of glare are vitreous debris and lenticular posterior capsular changes.[44] Bree et al. evaluated whether preoperative glare may affect the visual prognosis of patients with retinal dystrophies (RP, Usher syndrome type IIA, choroideremia, Alagille syndrome, and gyrate atrophy) after cataract extraction.[42] They calculated CDVA, spatial contrast sensitivity (SCS), temporal contrast sensitivity (TCS), and the measurable portion of glare (straylight) by C-Quant instrument before and after cataract surgery in 25 eyes of 16 patients. A statistically significant improvement was found for CDVA, SCS, and straylight, but TCS values did not change statistically significantly postoperatively. They considered functionally significant log(s) improvement as an increase of ≥0.20 log units and found that functionally significant improvement of straylight, SCS, and CDVA occurred in 72%, 32%, and 20% of eyes, respectively.

Postoperative improvement was not related to the preoperative value for both CDVA and SCS ($P = .33$ and $P = .51$, respectively). By contrast, postoperative straylight improvement was associated with the preoperative straylight value ($P < .05$). They observed a 50% chance of postoperative functionally significant log(s) improvement in eyes with a preoperative straylight value of 1.66 log(s); therefore, the preoperative straylight value can be used to predict a more likely postoperative improvement.

However, postoperative straylight improvement did not statistically correlate with postoperative CDVA improvement ($P = .56$) or with postoperative SCS improvement ($P = .13$).[42]

Finally, it is reasonable to consider the subjective report of symptoms as important as objective tests. Jackson et al. reported a 96.6% subjective improvement of symptoms, whereas the VA in 23% of eyes did not improve (unchanged or even worsened) postoperatively.[5] In other words, additional measures of visual function may be indicated when evaluating the *pros* and *cons* of cataract surgery in patients with RP, and further comprehensive studies are necessary to consider the subjective improvements.

Intraoperative Precautions

Phototoxicity As patients with only cataract and no retinal degenerative diseases are at risk for phototoxic retinal damage during surgery, patients with RP appear to be at greater risk.[5] Therefore, it is reasonable to reduce the intensity of illumination by 15% during the operation and also surgery time to limit macular damage.[5,9] Reducing the microscope light to 60% of normal intensity during the surgery and covering the light between different cataract steps have been suggested to minimize photic exposure.[35]

Zonular Insufficiency Zonular weakness is one of the most critical challenges in patients with RP, which occurs in 10% to 18.8% of cases.[4] It can result in complications such as intraoperatively vitreous loss and lens drop, postoperative anterior capsular contracture (ACC) syndrome (phimosis), and

Table 1. Effects of cataract extraction on patients with retinitis pigmentosa reported by previous studies

Authors, year, country	Patients (eyes) Age (y) Mean ± SD (range)	Follow-up (mo) Mean ± SD (range)	Study design	Preop CDVA (logMAR) Mean ± SD (range)	Postop CDVA (logMAR) Mean ± SD (range) P value	VA findings	Others
Jackson et al., 2001, UK[5]	89 (142) 47 (24-81)	32 (1-156)	Retrospective	1.05 ± 0.38 (0.3-0.2)	0.63 ± 0.49	Objective improvement in 77%, no change in 20.5%, and worsening in 2.5% Subjective improvement in 96%	
Figueroa-Wong et al., 2003, Mexico[36,a]	21 (28) 43	3	Retrospective			Improvement in 67.9% and no change in 32.1%	
Tokugawa et al., 2006, Japan[41,a]	16 (22) 61 (40-82)	3	Retrospective	0.14	0.42		No significant change in the visual field
Morimoto et al., 2007, Japan[38,a]	12 (21)		Retrospective			Improvement in 80.9%	
Wang et al., 2009, China[22,a]	21 (34)			<0.3 in 91.2%	<0.3 in 55.9% $P < .01$	Improvement in 91.2% and no change in 8.8%	Visual field improvement in 17.6% in the next 6 mo No significant change in ERG
Dikopf et al., 2013, USA[4]	47 (80) 48 (31-78)	23 (1 d-95 mo)	Retrospective	1.23	0.81 $P < .0001$	Significant improvement in most patients with RP with a preop vision of 20/200 or better Limited improvement in patients with a preop visual acuity of 20/400 or worse	
Garcia-Martin et al., 2013, Spain[21]	35 (35) 52 (45-66)	1	Cross-sectional	Snellen scale: 0.10 ± 0.23	Snellen scale: 0.48 ± 0.21 $P < .001$		Significant improvement in image repeatability after surgery
Takada et al., 2013, Japan[40,a]	7 (14) (39-71)			1.25 ± 0.71	1.11 ± 0.85	High rate of satisfaction after cataract surgery in patients with RP	
Bayyoud et al., 2013, Germany[33]	46 (52) 53 (21-89)	26 (3-60)	Retrospective	The entire group: 1.45 ± 0.85 Non-CTR group: 1.16 ± 0.8 CTR group: 1.74 ± 0.81	The entire group: 1.32 ± 0.95; $P = .02$ Non-CTR group: 0.98 ± 0.88; $P = .02$ CTR group: 1.66 ± 0.90; $P = .31$	Improvement in 52%, no change in 37%, and worsening in 12%	
Nakamura et al., 2014, Japan[39]	43 (58) (29-83)	3	Retrospective	0.81 ± 0.51	0.34 ± 0.43 $P < .0001$		The integrity of the IS/OS line may be important for predicting good postop CDVA

(continued on next page)

Table 1. Continued

Authors, year, country	Patients (eyes) Age (y) Mean ± SD (range)	Follow-up (mo) Mean ± SD (range)	Study design	Preop CDVA (logMAR) Mean ± SD (range)	Postop CDVA (logMAR) Mean ± SD (range) *P* value	VA findings	Others
Yoshida et al., 2015, Japan[9]	40 (56) 62 ± 10 (41-82)	37 ± 2 (6-73)	Retrospective	0.76 (0.08-2.30)	0.45 (0.18-2.00) *P* < .005	Significant improvement in the group with less advanced RP (MD was ≥−15 dB) No significant improvement in the group with advanced RP (MD <−15 dB)	No improvement of the final CDVA in 30 eyes (53.6%)
Davies et al., 2017, Canada[35]	18 (20) 52 ± 13	3 ± 3 (1 mo to several years)	Retrospective	1.09 ± 0.69 logMAR Snellen scale: 20/246 (hand motions to 20/25)	0.614 ± 0.448 logMAR Snellen scale: 20/82 (counting fingers at 2 feet to 20/20) *P* < .001	Significant improvement in both groups (preop visual acuity equal to or worse than 20/200 group and visual acuity 20/200 or worse group)	
De Rojas et al., 2017, USA[32]	(19) 51 ± 13	8.5 (3.5-30)	Retrospective cohort study	0.33 ± 0.20	0.19 ± 0.17	Improvement in 89% and no change in 11% Average improvement of 0.14 (SD 0.09) from baseline after cataract surgery	
Chan et al., 2017, China[34]	42 (67) 59 ± 12	82 ± 52	Retrospective	1.27 ± 0.42	1.18 ± 0.49 *P* = .095	55.2% of eyes returned to or worse than preop values	The mean duration of visual improvement after cataract surgery was 8.1 ± 0.8 y
Bree et al., 2017, Netherlands[42]	16 (25) 50 (28-71)		Prospective comparative study	0.36 ± 0.14	0.23 ± 0.14 *P* < .05	Straylight was the only parameter that could be used to support postop improvement	
Mao et al., 2018, China[37,a]	70 (109)	3	Retrospective	0.80	0.45 *P* < .001	Improvement in approximately half of the eyes	Preop CDVA and status of ELM and CMT are important parameters to predict postop VA

CMT = central macular thickness; CTR = capsular tension ring; ELM = external limiting membrane; ERG = electroretinography; IS/OS = inner and outer segments; MD = mean deviation; RP = retinitis pigmentosa
[a]Full texts were not available

intraocular lens (IOL) dislocation. Several methods have been reported and used to reduce zonular stress and complications, such as mechanical iris dilation (if needed), large capsulorhexis (about 6 mm), multiple radial relaxing incisions, gentle hydrodissection, extracapsular phacoemulsification, chopping technique, bimanual rotation of nucleus, viscodissection, implantation of 3-piece IOLs or large diameter 1-piece IOLs, and use of a capsular tension ring (CTR).[25,35] Because management of such complications may be complicated and need more surgical interventions, an alternative means of lens fixation or an anterior chamber IOL should be considered in patients with advanced zonular insufficiency.[4] A large capsulorhexis (6.0 mm or larger) size would help prevent the anterior capsular contraction in patients with RP; furthermore, implantation of IOLs with a larger optic diameter to maintain an adequate anterior capsular–optic overlap can decrease the risk of IOL tilt or decentration and also anterior and posterior capsular opacification.[45] Indeed, a considerable anterior capsular–optic overlap leads to extensive adhesion between the anterior capsule and the IOL optic, decreasing the migration of the anterior lens epithelial cells (LECs) behind the IOL optic.[46]

Capsular Tension Ring Dikopf et al. reported that only 5% of their cases required implantation of a 3-piece IOL rather than a standard 1-piece IOL, and no CTRs were implanted in their study.[4] Bayyoud et al. demonstrated that CTR implantation in such cases could cause fewer long-term complications such as posterior capsular opacity (PCO) and capsular contracture syndrome.[33] As a result, this method can be considered when necessary. However, some reports suggest that standard CTRs cannot prevent further zonular loss and, therefore, complications such as phimosis, lens subluxation, and dislocation of in-the-bag IOLs in progressive diseases such as RP.[47]

IOL Type It has been shown that selecting an appropriate IOL biomaterial and design could decrease the high incidence of PCO in patients with RP. Although poly(methyl methacrylate) (PMMA) and silicone implants carry a very high risk of PCO (approximately 90%) at 12 months of follow-up, acrylic-type IOLs may reduce the risk. In addition, it seems that hydrophobic material avoids the epithelial growth and fibrosis toward the visual axis center and, therefore, has superiority over hydrophilic material.[48] Vingolo et al. reported a significantly lower probability of postoperative PCO with the hydrophobic acrylic IOLs with a sharp-edged design in patients with RP compared with other types of IOL implants.[49]

Borkenstein et al. created hybrid monovision in a patient with RP for the first time, to our knowledge; they implanted a high-add lens (LENTIS MAX LS-313 MF80, Oculentis GmbH) in the right, near dominant eye and a monofocal, 7.0 mm optic IOL (ASPIRA-aXA, HumanOptics AG) in the left, far dominant eye.[45] Postoperatively, the patient's satisfaction was high; uncorrected distance and near VAs increased significantly. They suggested that using new IOLs and appropriate surgical planning, VA and especially the quality of life can be improved even in unpromising cases of incurable RP. However, considering the progressive course of RP and a lifelong risk of macular pathologies such as ERM or CME, the benefit of such approaches should be evaluated in larger studies with a longer duration of follow-up.

A number of surgeons prefer using blue light–filtering hydrophobic IOLs to reduce phototoxicity of blue light and the possibility of PCO formation.[9] By contrast, others believe that it is unclear whether blue light–filtering IOLs preserve macular health.[50] Akeo et al. reported an RP case for whom cataract surgery with implantation of colored IOLs was performed; retinal degeneration progressed, and color vision was severely impaired in this case.[51]

Anterior Lens Epithelial Polishing Dikopf et al. claimed that aggressive and consistent anterior lens epithelial polishing may result in mechanical disruption of the lens epithelial cells and could decrease the rate of anterior capsular phimosis and probably PCO in patients with RP.[4]

Femtosecond Laser–Assisted Cataract Surgery Johnson et al. reported a case of rapid and extreme capsular contraction after uneventful femtosecond laser–assisted cataract surgery (FLACS).[10] As the fellow eye, which had standard phacoemulsification with manual continuous curvilinear capsulorhexis (CCC) creation, did not show signs of anterior capsular phimosis, the authors suggested that femtosecond laser capsulotomy may be a risk factor for capsular contraction in patients with RP because of the release of inflammatory mediators from cell death during the laser capsulotomy combined with a structurally weaker CCC edge.[10] However, considering the potential advantages of FLACS in reducing the zonular stress and PCO formation, further clinical studies with larger samples should be performed in patients with RP.[52,53]

Postoperative Complications

Postoperative visits after cataract surgery in patients with RP should be conducted with greater detail and careful examination because they are at higher risk of complications compared with the normal population. Reported postcataract surgery complications in patients with RP are summarized in Table 2.[4,5,9,25,47,54–56] It was educational to search for the timing of complications (ie, mean and range of time) or any guidelines about scheduling follow-up visits in the literature; however, no relevant data were found, and

Table 2. Postcataract surgery complications in patients with retinitis pigmentosa

Complication	Studies	Prevalence	Prevention	Management
ACC	Yoshida et al.[9] Sudhir et al.[47]	Up to 25%	Using a CTR, creation of large capsulorhexis and radial relaxing incisions in the anterior capsule, and complete removal of lens epithelial cells	Nd:YAG laser
PCO	Yoshida et al.[9] Dikopf et al.[4] Jackson et al.[5]	Up to 85%	Implantation of foldable hydrophobic acrylic IOLs	Nd:YAG laser
Zonular weakness	Dikopf et al.[4]	Up to 20%		Use of a CTR, ECCE, viscodissection, bimanual rotation of the nucleus, gentle hydrodissection, and chopping techniques, IOL fixation methods, and AC IOL implantation
IOL dislocation	Hayashi et al.[55] Masket et al.[56],73 Najjar et al.[25] Sudhir et al.[47] Gimbel et al.[54]		Creating larger capsulorhexis, use of a CTR, and prevention of capsular phimosis	IOL exchange or fixation
CME	Dikopf et al.[4] Jackson et al.[5]	Up to 15%		Use of CAIs

ACC = anterior capsular contracture; CAI = carbonic anhydrase inhibitor; CME = cystoid macular edema; CTR = capsular tension ring; ECCE = extracapsular cataract extraction; PCO = posterior capsular opacity studies were focused on the prevalence of complications rather than the time of occurring postoperative complications.

Posterior Capsular Opacity Similar to capsular phimosis, disruption of the blood–ocular barrier and an increased level of proinflammatory materials such as IL-1 and IL-6 can lead to migration and proliferation of residual LECs and raised susceptibility of PCO formation.[57–60] Furthermore, the zonular insufficiency of these patients may result in a wrinkled posterior capsule, which can act as a scaffold for the migration of lens epithelial cells.[57] Previous studies reported a rate of 63% to 83.9% and 41.1% to 52.5% for PCO formation and performing YAG capsulotomy, respectively.[4,5,9]

The incidence of PCO in the normal eyes has been reported variably from less than 5% to 50% in different studies.[61] Many techniques have been examined and reported to prevent PCO in normal eyes or patients with RP.[48,62,63] These include interventions during surgery (eg, surgical techniques or IOL materials and designs) or pharmacological and nonpharmacological approaches interfering with the proliferation of LECs. Although some of these techniques, such as the implantation of acrylic IOLs with sharp-edged optics, are effective, widely accepted, and routinely used, they have not eradicated the complication of PCO.[62,64] On the other hand, performing a posterior CCC with or without bag-in-the-lens implantation of specially designed IOLs may entirely and permanently prevent significant PCO.[65,66]

Anterior Capsular Contracture A decrease in the anterior capsule opening after cataract surgery, known as ACC or capsular phimosis, usually happens in patients with diabetes mellitus, uveitis, pseudoexfoliation syndrome, and high myopia. It has been shown that the blood–retinal and blood–aqueous barriers of eyes with RP are defective.[66] After cataract surgery, such defects can lead to a raised level of cytokines in the aqueous humor and subsequent activity of lens epithelial cells. Lens epithelial cells show metaplastic changes and transform into myofibroblasts with the ability to form fibrous membranes.[60] Thus, fibrosis and contracture of the anterior capsule are expected complications in these eyes. Yoshida et al. reported an incidence of 23% for developing ACC in their study.[9] Creation of 6 mm capsulorhexis, polishing of the posterior capsule, cortical material, and anterior capsular epithelial cell removal as much as possible (with minimal trauma) were suggested techniques to prevent ACC.[67–69] It seems that the rate of developing phimosis is higher when using silicone IOLs.[70] However, an interesting case of severe capsular phimosis despite the application of CTRs and implantation of PMMA IOLs in a patient with RP has been reported.[47]

On the issue of comparison between FLACS and manual phacoemulsification, there is only 1 case report suggesting that severe inflammatory response and release of proinflammatory products after the femtosecond laser can accelerate the development of anterior capsular contracture.[10] This complication may cause decreased vision, IOL dislocation, and hypotony due to traction on ciliary processes.[71] Capsular phimosis can occur very soon, as in the case reported by Jin Poi et al. where phimosis presented 2 weeks after uneventful surgery, and the Nd:YAG laser can be used to manage this complication.[57,72] Early diagnosis and intervention is a crucial point and can prevent decentration, subluxation, and dislocation.[72]

IOL Dislocation RP is a commonly known risk factor for in-the-bag dislocation of IOLs.[55] Several mechanisms may play a role in this condition, including trauma during surgery or YAG laser capsulotomy, capsular phimosis, vitreous degeneration, and zonular insufficiency.[73] Creating a larger capsulorhexis and use of CTRs have been used to prevent capsular contracture and subsequent dislocation; however, the role of CTR is questionable because of numerous reports of IOLs and capsular complex dislocation despite the implantation of CTRs in progressive conditions such as RP and pseudoexfoliation syndrome.[47,71,73,74]

Although a CTR may have provided initial support, it is unknown whether this would have prevented or delayed late-onset decentration in these progressive cases.[75] However, in the event of postoperative CTR-IOL-capsular bag complex decentration, there are reports of successful and more easily accomplished surgical repositioning and scleral fixation with the presence of a CTR. In such cases, the CTR, acting as a backbone, may be directly sutured through the capsule to the sclera.[56,75]

Postoperative CME

In patients with RP, the prevalence of postcataract surgery macular edema or PCME detected on FA or OCT is about 10% to 14%, whereas this complication occurs approximately in only 1% to 4% of the general population.[4,5,9] Antonio-Aguirre et al., in a large retrospective multicenter cohort, demonstrated that the risk of postcataract surgery CME among patients with RP might be greater than 4 times that among the general population. They also showed that female sex and the presence of ERM (unlike in patients without RP) are associated with a lower risk of postoperative CME in eyes with RP.[76]

Topical carbonic anhydrase inhibitors (CAIs), oral CAIs, and local steroids are reported to be effective in treating RP-CME.[77] Still, regarding PCME in patients with RP, there is a lack of evidence or consensus on optimal treatment. The usefulness of CAIs in the treatment of PCME in patients with RP was reported in 1 case report.[78] Haruta et al. reported a case of a 63-year-old woman with RP who was initially diagnosed with CME and glaucoma. She underwent cataract surgery, vitrectomy, and sub-Tenon steroid injection 3 months later. At month 24, she still had persistent CME in the left eye. Based on fundoscopy, an electroretinogram was performed, and flattened waves led to the diagnosis of RP. Brinzolamide 1% was started in the left eye. Seven months later, on the Snellen chart, VA increased from 2/10 to 3/10, and on OCT imaging, CME improved. Hence, the authors concluded that CAIs seemed effective in treating PCME in patients with RP. The role of long-term postcataract medications in reducing the possibility of PCME is not clear yet. Davies et al. performed cataract surgery on 30 eyes from 18 patients with RP.[35] After the surgery, all eyes underwent a tapering course of topical steroid (prednisolone acetate 1%) and nonsteroidal anti-inflammatory drug (ketorolac 0.4%) over 3 months in addition to topical CAI (dorzolamide 2%) 2 times per day for 3 months; however, visually significant PCME (diagnosed on OCT) was still detected in 4 (13.3%) of 30 eyes.[35]

Similar Studies and Suggestions

Currently, 2 relatively similar comprehensive reviews are available in the literature about cataract surgery in patients with RP.[18,20] Our study had similar methods and results to the study by Hong et al. However, several important topics covered in our study have not been discussed in previous reviews, including (1) cataract surgical preferences of ophthalmologists in RP cases, (2) comparing functional and anatomical changes of the retina before and after cataract surgery through OCT, electroretinography, and VEP tests, (3) comparing visual parameters other than only VA, including glare, contrast sensitivity, and subjective symptoms, and (4) considering cases with preoperative CME. Also, this index study documented the beneficial effects of cataract extraction on patients with RP, including papers from 2001 to 2022 (compared with 2013 to 2018 in the study by Hong et al.) (Table 1).[18]

To date, cataract surgery treatment plans for patients with RP vary among clinicians based on their personal assessments. Providing a scoring system that considers functional and anatomical outcomes may pave the way for standardizing/unifying the approach to selecting eligible patients with RP for cataract surgery in the future. Further studies such as randomized clinical trials on visual and anatomical outcomes of these patients would also shed some light on this topic.

CONCLUSION

RP is the most common hereditary retinal degeneration that features progressive visual field loss. PSC is a common complication that occurs more frequently and earlier in patients with RP than in the general population. However, cataract extraction with IOL implantation in patients with RP is challenging. Surgeons should consider careful preoperative examination, proper case selection, intraoperative precautions, and postoperative management regarding probable complications. According to the literature, cataract surgery can lead to visual improvement in most cases even if the postoperative CDVA does not improve significantly, as patients report improvements in subjective vision. Possible suggested prognostic indicators to predict visual outcomes include preoperative CDVA (≥20/400), MD value in the visual field (≤−15), EZ integrity in OCT (grade 3), parafoveal flow density in the deep layer and superficial foveal avascular zone in OCTA, and straylight value (≥1.66). To reduce postoperative complications, several methods should be considered intraoperatively, such as mechanical iris retractors, large capsulorhexis, careful hydrodissection, chopping technique, use of a CTR, careful epithelial cell removal, and proper IOL selection. Postoperatively, regular and long-term follow-up is necessary for early detection and management of complications such as PCO, capsular contracture, IOL malposition, and macular edema.

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Disclosures: *None of the authors has any financial or proprietary interest in any material or method mentioned.*

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