Review Article

Cataract surgery in retinitis pigmentosa

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ABSTRACT

Background: Retinitis pigmentosa (RP) is an inherited retinal dystrophy characterized by progressive vision loss due to photoreceptor degeneration. Complicated cataract formation, particularly posterior subcapsular cataract (PSC), frequently occurs in RP and exacerbates the visual impairment. Cataract surgery may improve vision; however, the distinctive challenges of RP require specific considerations. This mini-review aims to provide a comprehensive overview of the RP-related cataract.

Methods: A comprehensive literature review was conducted via PubMed/MEDLINE, spanning the period from January 1976 to June 2024, using the keywords "cataract," "cataract surgery," "cystoid macular edema," "hereditary retinal dystrophy," "retinitis pigmentosa," "posterior subcapsular cataract," "posterior capsular opacification," "zonular weakness," and "artificial intelligence." We aimed to evaluate cataract surgery in patients with RP, focusing on cataract formation, its surgical management, postoperative complications, patient follow-up, and visual outcomes. Relevant review articles, clinical trials, and case reports with related reference lists of these articles were included.

Results: A total of 53 articles were examined in detail, including those identified through focused keyword searches and the reference lists of these articles. Cataract surgery in patients with RP generally results in substantial visual improvement. However, surgery can be complicated, particularly by zonular weakness and subluxation of the crystalline lens. These risks can be reduced by using capsular tension rings and employing meticulous surgical technique. Furthermore, postoperative complications, such as cystoid macular edema and posterior capsular opacification, are common. Despite these challenges, regular postoperative follow-up and appropriate management can help mitigate complications. Integrity of the ellipsoid zone and external limiting membrane on preoperative optical coherence tomographic examination are the main predictors of visual outcomes following cataract surgery; however, outcomes can vary. Though many patients experience significant visual improvement, some may experience limited benefits due to pre-existing advanced retinal degeneration.

Conclusions: Cataract surgery may offer meaningful visual benefits in patients with RP; however, careful preoperative evaluation and meticulous surgical technique are required to address the possible challenges. Attentive postoperative care and follow-up are essential to optimize visual outcomes. Early surgical intervention can significantly improve the quality of life in selected candidates, and tailored approaches are necessary in patients with RP requiring cataract surgery. Further studies on the potential application of artificial intelligence to monitor postoperative recovery and detect complications may improve surgical outcomes and enhance patient care.

KEYWORDS

cataracts, cataract extractions, cystoid macular edema, retinitis pigmentosa, tapetoretinal degeneration, posterior subcapsular cataract, capsule opacifications, zonular weakness, computational intelligence, AI (artificial intelligence)

INTRODUCTION

Retinitis pigmentosa (RP) is the most common form of inherited retinal dystrophy. It is an umbrella term for a diverse group of inherited retinal diseases that initially cause degeneration of rod photoreceptors, followed by the cone photoreceptors [1, 2]. This progressive degeneration is a leading cause of visual impairment and blindness in people younger than 60 years, affecting approximately 1 in 4000 people worldwide [1-3].

The genetic mutations that cause RP result in specific biochemical dysfunctions that affect the rod photoreceptors in the retina. RP is genetically heterogeneous, implying that the disease is associated with numerous genetic mutations [4, 5]. These mutations impact genes that are critical for retinal functions, including phototransduction, visual cycle, and photoreceptor structural integrity. More than 80 identified genes contribute to the diverse patterns of inheritance and manifestations observed in RP [4, 6, 7].

The diagnosis of RP is primarily based on clinical findings, such as night blindness, peripheral vision loss, and prominent retinal changes confirmed by abnormal electroretinogram results. Typical fundus changes include bone spicule hyperpigmentation and hypopigmentation, waxy disc pallor, and arteriolar narrowing [8]. Other fundus features include optic disc drusen, vitreous cells, epiretinal membranes, cystoid macular edema (CME), and exudative vasculopathy resembling Coats' disease [4, 8]. Findings of a typical patient with RP and CME are shown in Figure 1.

Along with these fundus changes, cataract is the most common complication in patients with RP, often contributing to vision impairment [9]. Posterior subcapsular cataract (PSC) is the most common type of cataract in patients with RP, followed by nuclear cataract (NC) [10]. The prevalence of cataract varies among studies conducted in different populations. Pruett [11] reported that cataracts were observed in 46.4% of eyes with RP, of which 93.6% were PSC. Similarly, the frequency of PSC was found to be 41% by Merin et al. [12] and 44.4% by Fujiwara et al. [9]. Fishman et al. [13] identified PSC in 180 (53%) of 338 patients with RP. Central PSC with a clear nucleus typically occurs in the middle stages of disease progression, and the severity of lens opacity may increase with age or disease duration [10, 14].

In patients with RP, cataract surgery is usually required between the ages of 47 to 58 years, whereas in the general population with age-related cataracts, it is usually required between the ages of 72 to 74 years [15]. Approximately 2% of individuals aged between 52 and 85 years have PSC. In contrast, 50% of individuals with RP develop PSC earlier than those with age-related cataracts [13, 15]. NC is less common than PSC and occurs at an average of 20 years later than PSC in patients with RP [16]. Lens opacities typically appear in the middle stages of the disease and are associated with noticeable clinical symptoms and signs. Because of the pre-existing constriction of visual field in patients with RP, even small lens opacities can lead to a disproportionate reduction in visual acuity (VA). Thus, patients often require cataract surgery earlier than those with age-related cataracts [10].

This mini-review aims to provide a comprehensive overview of RP-related cataract, focusing on cataract surgery, associated complications, patient follow-up, and visual outcomes.
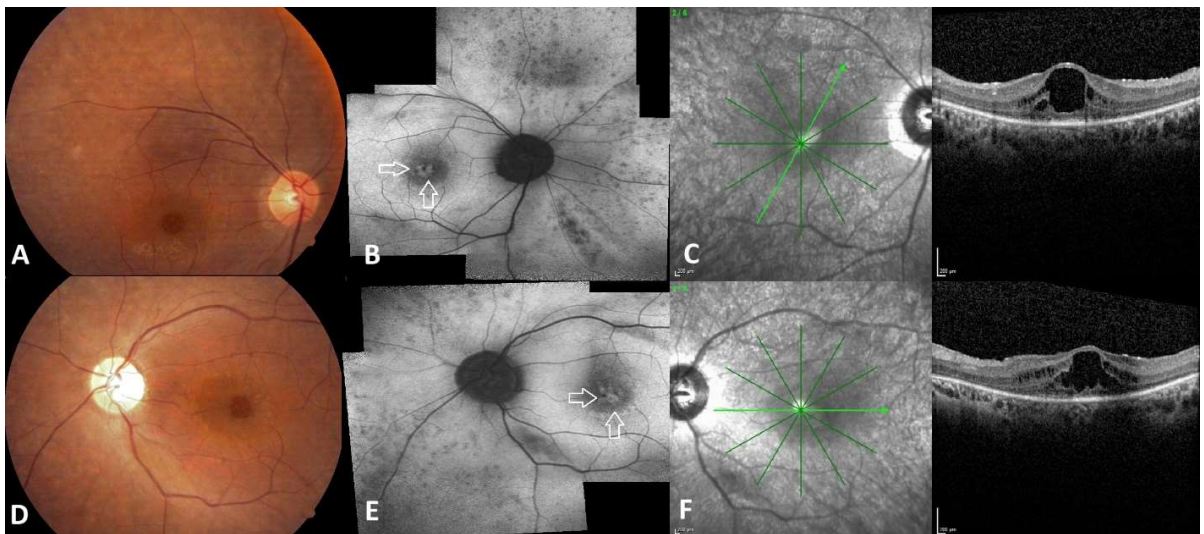

Figure 1. Color fundus images of the right (A) and left (D) eyes, displaying the attenuated blood vessels and slightly pallid left optic nerve. Fundus autofluorescent images of the right (B) and left (E) eyes, exhibiting multiple patchy hyperautofluorescent and hypoautofluorescent areas corresponding to the pigment deposits and areas of retinal pigment epithelial atrophy. In addition, there is a bilateral petaloid hyperautofluorescence pattern (white arrows) compatible with cystoid macular edema. Optical coherence tomographic images of the right (C) and left (F) eyes depict the presence of cystoid changes with disrupted external limiting membrane and ellipsoid zone.

METHODS

A comprehensive literature search was conducted using PubMed/MEDLINE, spanning the period from January 1976 to June 2024, using the keywords "cataract," "cataract surgery," "cystoid macular edema," "hereditary retinal dystrophy," "retinitis pigmentosa," "posterior subcapsular cataract," "posterior capsular opacification," "zonular weakness," and "artificial intelligence." This search aimed to identify studies evaluating cataract development, management, and the outcomes of cataract surgery in patients with RP. Additionally, the reference lists of included studies were reviewed.

RESULTS

A total of 53 articles were examined in detail, including those identified through focused keyword searches and the reference lists of these articles. Included papers were comprehensively reviewed by two senior ophthalmologists with retina subspecialty to explore the pathogenesis of complicated cataract in patients with RP, the various dimensions of cataract surgery in patients with RP, cataract surgery-related postoperative complications, postoperative follow-up, and visual outcomes.

Table 1. Key findings from major studies on outcomes of cataract surgery in patients with RP

Author (Year)	Study design and sample size	Key findings
Jackson et al. (2001) [42]	Retrospective 89 patients (142 eyes)	Mean age at surgery → 47.5 years (range, 24–81 years). Cataract type → PSC alone (100 eyes), PSC and NC (37 eyes), and NC (5 eyes). Mean ± SD VA (logMAR) improved from 1.05 ± 0.38 to 0.63 ± 0.49 after surgery. Changes in VA → Improved in 109 eyes (77%), unchanged in 29 eyes (20.5%), and worsened in 4 eyes (2.5%). The rate of PCO → 63% (88/139 eyes), and 45.1% required Nd:YAG laser capsulotomy. The rate of postoperative CME → 14% (20 eyes).
Dikopf et al. (2013) [26]	Retrospective observational 47 patients (80 eyes)	Mean follow-up time → 23.3 months (range, 1 day–95 months). The rate of PSC → 97.5%. The mean BCVA (logMAR) significantly improved post-cataract surgery → from 20/340 to 20/129 within 3 months of surgery ($P < 0.0001$). The rate of PCO → 82.5% (66 eyes), and 52.5% (42 eyes) required Nd:YAG laser capsulotomy at an average of 10.8 months after surgery. The signs of phacodonesis → 15 eyes (18.8%) of 10 patients (21.3%) (preoperatively → 3 eyes, intraoperatively → 8 eyes, and postoperatively → 4 eyes). One patient experienced bilateral dislocation of in-the-bag intraocular lenses at 5.5 years and 6 years post-surgery.
Yoshida et al. (2015) [47]	Retrospective observational 40 patients (56 eyes)	The mean BCVA (logMAR) significantly improved post-cataract surgery, from 0.76 to 0.45 ($P < 0.005$). BCVA did not improve in 30 eyes (53.6%). The final BCVA was notably better in patients with a normal EZ on preoperative OCT compared to those with an abnormal or invisible EZ. The preoperative MD value on the HFA 10-2 program and the presence of a normal inner/outer segment on OCT significantly influenced the final BCVA. The rate of PCO → 83.9% (47 eyes), and 41.1% (23 eyes) required Nd:YAG laser capsulotomy over a follow-up period of 3 years.
Nakamura et al. (2015) [46]	Retrospective observational 64 patients (96 eyes)	The mean ± SD follow-up period → 5.8 ± 2.4 years. The mean ± SD BCVA (logMAR) → Preoperative → 0.64 ± 0.52, and final postoperative → 0.61 ± 0.67 ($P = 0.57$). A significant improvement in postoperative BCVA was observed only in eyes with intact foveal EZ ($P < 0.01$). Significantly lower MD, macular sensitivity, and foveal sensitivity at the final visit in 62 eyes of 45 patients when compared to preoperative measurements ($P < 0.01$).
Mao et al. (2018) [30]	Retrospective observational 70 patients (109 eyes)	A significant improvement in mean BCVA (logMAR) after cataract surgery, from 0.80 to 0.45 ($P < 0.001$). Final BCVA did not improve in 57 eyes (52.3%). Postoperative BCVA was significantly better in eyes with an intact ELM compared to those without, in eyes with an intact EZ compared to those without, and in eyes with relatively normal CMT (≥ 200 μm) compared to those with reduced CMT (< 200 μm). Three variables were significantly associated with postoperative BCVA → Preoperative logMAR VA ($P < 0.001$), ELM integrity ($P < 0.001$), and CMT ($P = 0.017$).
Nguyen et al. (2023) [41]	Retrospective 226 patients (295 eyes)	The mean ± SD age of patients undergoing surgery → 56.1 ± 17.9 years. Significant BCVA improvements in most patients after surgery. Greater odds for marked visual improvements in patients with moderate or worse visual impairment. The most frequently encountered complications → PCO (n = 111, 38%), zonular dialysis (n = 15, 5%), and CME (n = 14, 5%).
Sakai et al. (2023) [48]	Retrospective 38 patients (38 eyes)	The median BCVA (logMAR) significantly improved after cataract surgery. Median BCVA (logMAR) improved from 0.52 to 0.07. Postoperative BCVA showed a significant correlation with the horizontal (r = - 0.784; $P < 0.001$), vertical (r = - 0.777; $P < 0.001$), and average (r = - 0.777; $P < 0.001$) EZ widths.
Georgiou et al. (2024) [45]	Retrospective comparative 52 patients (72 eyes) in RP group 83 468 patients (113 317 eyes) in control group	Worse preoperative VA (mean logMAR = 1.03 versus 0.59, $P < 0.001$) and postoperative VA (0.71 versus 0.14, $P < 0.001$) in RP group compared to control group. The mean ± SD VA gain → 0.25 ± 0.60 logMAR in RP versus 0.43 ± 0.48 logMAR in control group ($P < 0.001$). No significant differences in the rate of intraoperative pupil expansion use, posterior capsular tears, dropped nucleus, or zonular dialysis between the groups. Postoperative CME was significantly more common in eyes with RP (6.9%) compared to the control group (1%) ($P < 0.001$). No significant difference between the two groups regarding the need for IOL repositioning or exchange. The rate of Nd:YAG capsulotomy → 13.9% in RP and 3% in control group.

Abbreviations: RP, retinitis pigmentosa; PSC, posterior subcapsular cataract; NC, nuclear cataract; VA, visual acuity; SD, standard deviation; PCO, posterior capsular opacity; CME, cystoid macular edema; BCVA, best corrected visual acuity; EZ, ellipsoid zone; ELM, external limiting membrane; CMT, central macular thickness; IOL, intraocular lens; MD, mean deviation; HFA, humphrey field analyzer; logMAR, logarithm of the minimum angle of resolution; OCT, optical coherence tomography.

DISCUSSION

Pathogenesis of complicated cataract in RP

The exact mechanism underlying cataract formation in patients with RP remains incompletely understood. The inflammatory response is a substantial aspect of RP [16] and is characterized by activation of the innate immune system, including breakdown of the immune barrier, activation and infiltration of immune cells, and increased expression of both local and systemic inflammatory factors. In addition, breakdown of the blood–retinal barrier often results in the presence of inflammatory cells in the vitreous [17, 18]. Microglia, a critical feature of the retinal innate immune system, play a key role in retinal inflammatory processes [19]. Several studies have reported increased levels of inflammatory factors (e. g., interleukin [IL]-1, IL-2, IL-4, IL-6, monocyte chemoattractant protein-1, tumor necrosis factor-alpha [TNF-α], and placental growth factor) in the serum, vitreous, and aqueous humor, implying an active inflammatory response in patients with RP [17, 20, 21]. The intraocular microenvironment is altered by the inflammatory response associated with RP [10], with changes similar to those observed in cataracts associated with uveitis [22]. The inflammation-induced alteration of the microenvironment is critical in the development of cataracts in patients with RP [10].

The ultrastructural formation of PSCs in the Royal College of Surgeons rat, an animal model of human autosomal recessive RP, has been demonstrated using scanning electron microscopy (SEM), transmission electron microscopy (TEM), and light microscopy (LM) [23]. In these rats, retinal degeneration occurred from 2 to 6 weeks after birth, leading to development of PSCs [23]. The researchers proposed that this was likely due to toxic lipid peroxides produced by degenerating rod outer segments. They also demonstrated that these PSCs were morphologically characterized by the proliferation of dysplastic bladder-like fibers, or Wedl cells, in the meridional region of the lens. These cells eventually migrate and accumulate at the posterior pole of the lens to form the PSC [23]. In another study of patients with RP, Andjelic et al. [15] examined the anterior capsule obtained during cataract surgery using SEM and TEM. They identified several abnormal features in the anterior lens epithelium, including holes, thinning, epithelial degradation, and cracks between the adjacent lens epithelial cells. The authors proposed that these structural irregularities could facilitate water influx into the lens, potentially leading to opacification along the water clefts extending toward the posterior pole. They believed that the lens epithelium played an important role in the development of cataracts in patients with RP [15].

Management of cataract surgery in RP

In addition to the progressive degeneration of photoreceptors and retinal pigment epithelium, cataract formation can further compromise the remaining useful vision [24]. Even if the cataract is not severe, its central location can still cause blurriness in the remaining central visual field. In patients with RP, even mild lens opacity can result in significant functional impairment. Therefore, cataract surgery with intraocular lens (IOL) implantation may become a requisite [14]. However, the necessity of cataract surgery in patients with RP is controversial, as some believe that the procedure may accelerate photoreceptor degeneration by light exposure, potentially worsening the VA and leading to postoperative complications such as posterior capsule opacification (PCO) [24, 25]. Thus, the decision for surgery and its timing involves tailored management strategies in patients with RP. The following key issues should always be considered:

Preoperative assessment: Comprehensive evaluation of the patient's ocular health is mandatory in the preoperative assessment, including best-corrected visual acuity (BCVA) and stability of the crystalline lens [10]. Phacodonesis with zonular insufficiency is substantially more common in patients with RP [10, 26-28]. Dikopf et al. [26] reported a phacodonesis rate of 18.8% (15 of 80 eyes) in patients with RP. However, preoperative detection of phacodonesis was possible in only about 20% of these cases. Zonular weakness and dehiscence in patients with RP have been attributed to early vitreous liquefaction, irregular vitreous scaffolds, and toxic substances associated with long-term inflammation. Identifying zonular insufficiency is crucial for surgical planning, technique selection, and preventing potential intraoperative and postoperative complications [10, 26-28].

In addition to anterior segment evaluation, retinal evaluation using optical coherence tomography (OCT) is crucial prior to cataract surgery in patients with RP. Preoperative central macular thickness (CMT) and integrity of the external limiting membrane (ELM) and ellipsoid zone (EZ) are significant predictors of postoperative visual outcome. Assessing these retinal layers can help clinicians predict the improvement in VA and guide the surgical approach [24, 29, 30]. CME is a potential complication in RP [31], affecting 10% to 50% of patients. Degeneration of the retina and retinal pigment epithelium is believed to release "toxic products" that lead to RP-related CME by disrupting the blood–retinal barrier [32]. Similar to CME in other diseases, RP-related CME has been associated with the release of several factors, including vascular endothelial growth factor (VEGF), TNF-α, IL-1α, IL-1β, adenosine, prostaglandins, histamine, and insulin-like growth factor [32]. Interestingly, RP-related CME may not always correlate with a reduction in VA [32, 33]. Oishi et al. [34] found no correlation between total macular thickness and VA. However, the integrity of the EZ has been shown to correlate well with VA and may provide information about the visual prognosis. De Rojas et al. [29] reported that none of the patients with RP and preoperative CME experienced a postoperative decrease in VA [29]. However, recognizing CME prior to surgery is crucial for setting realistic postoperative visual expectations for both the patient and the surgeon [35].

Surgical management: Considering the unique challenges of RP, phacoemulsification with IOL implantation is the preferred surgical method. Pre-existing zonular weakness in some patients renders the intraoperative maintenance of zonular integrity crucial to prevent zonular insufficiency-related problems [10, 24]. In patients with zonular weakness, capsular tension rings (CTRs) should be inserted intraoperatively to support the zonules and stabilize the IOL postoperatively [10]. Moreover, in some cases, surgeons might prefer three-piece IOL designs, as they provide more possibilities for suture fixation if the IOL–bag complex becomes dislocated in the future [36]. In a retrospective study, Bayyoud et al. [37] evaluated the long-term postoperative outcomes with a mean follow-up of 26 months after cataract surgery in patients with RP. They compared the results between those who received CTR implantation and those who did not. The utilization of CTRs during cataract surgery was associated with a lower incidence of long-term complications, including PCO formation and capsular contraction syndrome (CCS) [37]. Similarly, Chen et al. [38] reported that none of the patients with RP who underwent CTR implantation developed postoperative CCS. Therefore, the authors emphasized that the use of CTRs reduced complications due to capsular contraction [38].

Another critical consideration in RP surgery is the choice of IOL biomaterial. In a retrospective study by Bruno et al. [39], three types of IOL biomaterials—polymethyl methacrylate (PMMA), silicone, and acrylic—were compared in patients with RP to evaluate the incidence of postoperative PCO. The study found that 89.9% of patients with PMMA IOLs and 90.9% of patients with silicone IOLs required Nd:YAG capsulotomy at 12 months, compared to only 52.8% of patients with acrylic IOLs. Additionally, when comparing acrylic materials, the authors found that 41.3% of patients with hydrophobic acrylic IOLs developed PCO, whereas 60.0% of patients with hydrophilic acrylic IOLs experienced this complication. Therefore, the authors suggested that the smooth surface of hydrophobic acrylic lenses helps prevent obscuration of the central visual axis by epithelial cell growth and fibrosis [39]. In contrast, there have been no publications in the literature regarding the use of extended depth of focus (EDOF) IOLs and multifocal IOLs in RP. Currently, multifocal IOLs should be strictly avoided in patients with RP, as they may reduce contrast sensitivity [40].

Postoperative care, complications, and management: Although cataract surgery can be visually beneficial in patients with RP, it may also be fraught with complications, such as intraoperative phototoxic retinal damage and postoperative pseudophakic CME, PCO, CCS, and IOL subluxation or dislocation [10, 24, 26, 37-39, 41].

Pseudophakic CME is a common complication after cataract surgery in patients with RP. Studies have shown that the risk of developing CME related to cataract surgery is more than four times higher in patients with RP compared to those without RP. The incidence of macular edema or pseudophakic CME in patients with RP ranges from 13.3% to 32%, whereas in the general population, this complication occurs in approximately 1.1% to 4.2% of cases following cataract surgery [10, 24, 26, 35, 41-44]. The increased incidence of CME in patients with RP underscores the need for thorough preoperative and postoperative evaluation to minimize risk. The use of OCT to detect pre-existing CME before surgery can help inform patients of the potential for worsening CME and allow treatment to be initiated before the procedure [45]. The use of topical nonsteroidal anti-inflammatory drugs (NSAIDs), either in conjunction with or as an alternative to topical corticosteroids, can markedly reduce the risk of developing CME after cataract surgery. In high-risk patients, prophylactic treatment should ideally begin a few days prior to surgery and continue for at least three months postoperatively [43]. Although acute CME often resolves spontaneously, prompt diagnosis and appropriate treatment are crucial to prevent retinal damage and permanent visual impairment [43]. Topical NSAIDs, carbonic anhydrase inhibitors (CAIs), and corticosteroids are effective treatments for acute pseudophakic CME, whereas oral CAIs, periocular and intravitreal steroids, and intravitreal anti-VEGF agents can be used for refractory cases [43, 45].

PCO is the most common sight-threatening complication after cataract surgery and is more aggressive in eyes with RP compared to healthy eyes. PCO results from capsular fibrosis driven by lens epithelial cell migration, proliferation, and epithelial–mesenchymal transition, which may be exacerbated by elevated cytokine levels in eyes with RP [10, 35]. The rate of PCO formation ranges from 63% to 83.9%, whereas the rate of performing Nd:YAG capsulotomy ranges from 41.1% to 52.5% [26, 35, 42, 46]. The incidence of PCO is higher in younger patients, likely due to differences in wound healing responses. Because most cataract surgery in patients with RP occurs at a younger age, this may partially explain the increased risk of PCO in this population [10]. Patients with RP who develop PCO can be effectively treated using a Nd:YAG laser [10].

RP is widely recognized as a risk factor for in-the-bag dislocation of IOLs. Several mechanisms can contribute to this condition, including surgical trauma, Nd:YAG laser capsulotomy, capsular phimosis, vitreous degeneration, and zonular insufficiency. To reduce the risk of capsular contracture and subsequent dislocation, the creation of a larger capsulorhexis and the use of CTRs have been recommended [35]. Surgery is the primary treatment for these patients, and several techniques have been developed to reposition the subluxated or dislocated IOL with suture or sutureless techniques [36].

Follow-up and visual outcome: In many patients with RP, cataract surgery can significantly improve VA and overall quality of life. However, progressive photoreceptor degeneration, postoperative complications, postoperative inflammatory responses, and retinal damage from light exposure—in which photopigments, retinoids, and bisretinoids contribute to photochemical damage—have led to mixed conclusions regarding the long-term effects of cataract surgery in patients with RP [24]. In addition, not all patients with RP are ideal candidates for cataract surgery, owing to poor preoperative retinal conditions, which may limit the improvement in VA [24]. The main studies reporting outcomes of cataract surgery in patients with RP are discussed below and summarized in Table 1 [26, 30, 41, 42, 45-48].

Yoshida et al. [47] reported that approximately half of the eyes with RP experienced improved final BCVA after cataract surgery. Similarly, Nguyen et al. [41] reported that significant visual improvements were observed in 87 of 226 patients with RP (39%) following cataract surgery. They found that patients with moderate to severe visual impairment were more likely to achieve significant visual improvement and suggested that baseline BCVA was a good predictor of the visual outcome [41]. In a recent comparative study by Georgiou et al. [45], the authors evaluated the visual outcomes of isolated cataract surgery in eyes with RP compared to non-RP eyes. While VA improved postoperatively in patients with RP, both preoperative and postoperative VA were worse in the RP group, and the mean VA gain was significantly lower in these patients. The authors concluded that cataract surgery could improve vision in eyes with both RP and cataract [45].
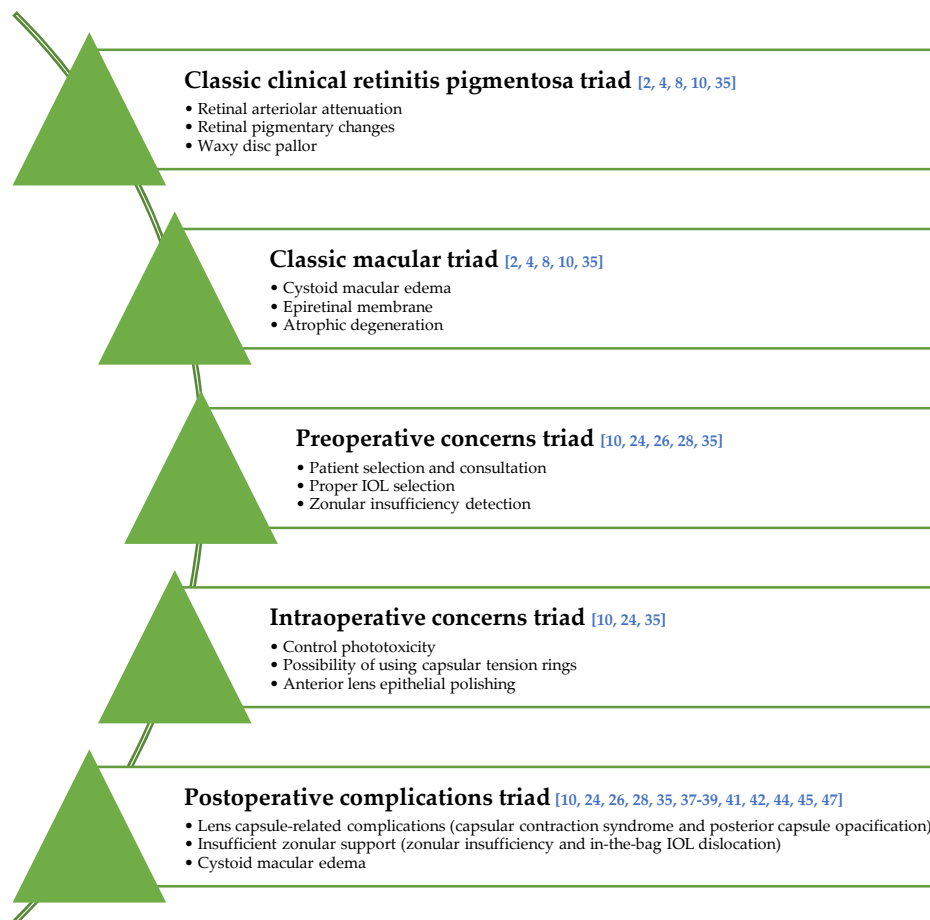
In a systematic meta-analysis by He et al. [24], the authors reported that cataract surgery could improve VA in patients with RP at different follow-up periods. They indicated that postoperative VA could be improved by 0.57 log units compared to preoperative values. However, the VA improvement showed a reverse trend after more than 1 year of follow-up. The authors suggested that this might be caused by PCO formation typically between 6 months and 1 year after surgery. They also used OCT to evaluate visual outcomes after cataract surgery in patients with RP and different degrees of preoperative macular EZ integrity. Their results showed no significant difference between preoperative and postoperative VA in the EZ-invisible group. Therefore, they recommended against cataract surgery in patients with EZ-invisible RP, as no visual improvement could be anticipated after surgery. The authors also recommended that OCT should always be performed in patients with RP prior to cataract surgery [24]. Nakamura et al. [46] reported similar findings, emphasizing that a normal IS/OS line on OCT examination was associated with a favorable visual recovery after cataract surgery in patients with RP. They also emphasized that integrity of the IS/OS line might be critical in predicting a good postoperative BCVA [46]. In another observational study by Mao et al. [30], the authors emphasized that preoperative BCVA, along with the status of the ELM and CMT, were critical predictors of postoperative VA [30].

A key aspect of the cataract surgery debate is whether the procedure accelerates disease progression. To address this question, De Rojas et al. [29] investigated the potential impact of cataract surgery on the progression of RP. The study included 70 eyes of 40 patients with RP, divided into two groups: those who underwent phacoemulsification with IOL implantation and those who did not undergo cataract surgery. OCT was used to assess the width of the EZ, a reliable indicator of RP severity, at baseline and during follow-up (median 768 days). RP progression was determined by measuring the loss of EZ width over time for all participants. A multivariable analysis adjusting for factors such as age, baseline EZ width, mode of inheritance, and cataract surgery status showed no significant difference in disease progression between the surgery and control groups. VA improved in almost all eyes that underwent surgery (17 of 19 [89%]), whereas it remained stable in the remaining eyes (2 of 19 [11%]). Therefore, the authors concluded that cataract surgery is a safe and effective method to improve VA in patients with RP, and that it does not appear to accelerate disease progression as assessed by OCT [29]. In light of the above information, a brief summary of RP and the management of cataract surgery is presented in Figure 2.

Artificial intelligence (AI) has become a promising tool in ophthalmology, revolutionizing the diagnosis and management of eye diseases. AI has enabled the early diagnosis of RP and the prediction of visual function in patients with RP [49-53]. In future studies, AI algorithms may be employed to analyze imaging data with the objective of identifying the early formation of cataracts, forecasting their progression, and tailoring surgical plans to align with unique individual patient profiles. Furthermore, AI may have the potential to monitor the postoperative recovery and detect complications at an early stage, resulting in improved surgical outcomes and enhanced patient care.

This review has both strengths and limitations. The main strength is its nearly five-decade review, which offers a profound insight into the evolution of surgical techniques and related outcomes. The major limitations include heterogeneity of design in the included studies and the predominance of retrospective studies. Further research should comprise prospective studies of the long-term consequences and underlying genetics and molecular mechanisms involved in cataract formation in eyes with RP. Additionally, the development of novel surgical techniques customized to this specific patient cohort could be a fruitful avenue for investigation. Further comprehensive reviews using a systematic review and meta-analytic strategy with additional databases may shed more light on the subject.

Figure 2. Triads of diagnostic findings along with pre-, intra-, and postoperative issues in eyes with retinitis pigmentosa.

Classic clinical retinitis pigmentosa triad [2, 4, 8, 10, 35]
• Retinal arteriolar attenuation
• Retinal pigmentary changes
• Waxy disc pallor

Classic macular triad [2, 4, 8, 10, 35]
• Cystoid macular edema
• Epiretinal membrane
• Atrophic degeneration

Preoperative concerns triad [10, 24, 26, 28, 35]
• Patient selection and consultation
• Proper IOL selection
• Zonular insufficiency detection

Intraoperative concerns triad [10, 24, 35]
• Control phototoxicity
• Possibility of using capsular tension rings
• Anterior lens epithelial polishing

Postoperative complications triad [10, 24, 26, 28, 35, 37-39, 41, 42, 44, 45, 47]
• Lens capsule-related complications (capsular contraction syndrome and posterior capsule opacification)
• Insufficient zonular support (zonular insufficiency and in-the-bag IOL dislocation)
• Cystoid macular edema

CONCLUSIONS

Cataract surgery in patients with RP is a viable intervention that can significantly improve VA and quality of life. Current findings suggest that surgery does not affect the progression of RP. Despite the inherent complexity and higher risk of complications, such as CME, PCO, and zonular insufficiency, advances in surgical techniques and preoperative assessment have improved outcomes. Assessing the integrity of the EZ and ELM, along with careful management of intraoperative and postoperative complications, is critical in predicting and optimizing visual recovery. Overall, with meticulous preoperative assessment and tailored surgical techniques, cataract surgery is visually beneficial in patients having RP with cataracts.

ETHICAL DECLARATIONS

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