Optical Iridectomy as An Alternative Clear Visual Axis for Peters Anomaly

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ABSTRACT

Introduction and objective: Peters anomaly is a rare congenital disease presented with central leukoma, iridocorneal adhesion, and with or without a cataractous lens. The presence of central leukoma will block the visual axis and lead to a disruption in normal visual development. Therefore, optical iridectomy, which can overcome the high incidence of graft failure in penetrating keratoplasty (PK), has been proposed as the alternative treatment to clear the visual axis. This study aims to show the outcome of optical iridectomy in patients with Peters’ anomaly.

Method: Systematic literature searched in Pubmed, Google Scholar, and Cochrane from their inception to August 2020 was conducted using keywords “iridectomy” and “Peters’ anomaly”. Full-text articles in English that report the outcome of iridectomy in Peters’ anomaly were included, and a manual review of article bibliographies was done. Outcome measures were analyzed using the following clinical questions: indication, age, visual acuity (VA), and intraocular pressure (IOP).

Result: One case series and two case reports demonstrating the use of optical iridectomy in Peters anomaly were identified. In total, 26 patients aged one week to 7.7 years received optical iridectomy to establish a clear visual axis. All studies showed an improvement in visual acuity through clinical findings, and one of the studies showed a better postoperative VA in bilateral cases. Studies showed good control of IOP.

Conclusion: Optical iridectomy can be used as a safe procedure to improve visual acuity in Peters anomaly patient.

Keywords: anterior segment dysgenesis, optical iridectomy, peters anomaly

Peters anomaly is one of the congenital anomalies that affect the anterior segment of the eye resulting in central cornea opacities and other anterior segment disruption.¹ Central corneal opacities will block the visual axis which in turn will disrupt the normal visual development. Thus, immediate treatment is necessary to prevent sensory ambliopia. To this day, the most commonly performed surgery to treat the central leukoma is penetrating keratoplasty.² However, this technique still has several shortfalls, and the most concerning one is the high incidence of graft failure.¹³⁵ Therefore, optical iridectomy was proposed as an alternative treatment. The aims of this study is to know the outcome and complication of optical iridectomy in patients with Peters anomaly.

METHODS

Literature search was conducted through Pubmed, Google Scholar, and Cochrane from their inception to August
2020 using keywords “iridectomy” and “Peters anomaly”. Randomized control trials, cohort studies, case reports, and case series related to the outcome of optical iridectomy in patients with Peters anomaly were retrieved. Article bibliographies were manually reviewed to find the other relevant studies.

The abstract and full-text studies in English were evaluated, and reports that did not fulfill the objective of the study were excluded. The data of study design, indication, and age were gathered as demographic data, while visual acuity and intraocular pressure were included as a marker of improvement. Any adverse effects related to the procedure were also noted.

RESULT

Three articles in English illustrating the outcome of optical iridectomy in Peters anomaly were identified. In total, 35 eyes from 26 patients underwent optical iridectomy to treat their leucoma. The age of patients was ranged from one week to 7.7 years (Table 1). Out of the total articles, two case series, and one case report, all showed the same indication, which is Peters’ anomaly type I with a clear peripheral cornea.

A study by Zaidmann et al. was done in three patients, ranging from 10 weeks old to five months old, who had paracentral corneal opacities. Two of those had undergone trabeculectomy prior to optical iridectomy. At the last follow-up, all patients maintained a clear visual axis, as the examiner can have a good view of patients’ fundus. However, the ability to fixate and follow was only reported in two patients. All patients showed a normal IOP post-operatively and healed without any complications. Nevertheless, those with previous glaucoma surgery still used their glaucoma medications.

The second study, which is the largest study was done by Spierer et al. in 2018. The study included 22 patients (29 eyes), and the mean age at the time of surgery was 15.6 ± 26.3 months. The opacity involved the central visual axis, and no red-reflex was observed preoperatively in all of the patients. Optical iridectomy was most commonly performed in the temporal area with a minimum three-hour-clock sized. Postoperatively, 98.6% (28 eyes) showed a red reflex. One patient who did not show a red reflex undergoes another temporal iridectomy. At the long-term follow-up, around eight months to 72 months post-surgery, three of those who had a red reflex need to undergo another surgery. Two of them having penetrating keratoplasty, and one of them having keratoprosthesis. The improvement in the mean visual acuity (VA) was statistically significant (p<0.001). Furthermore, the postoperative visual acuity improved significantly on bilateral disease (p<0.05). However, three eyes (10.3%) showed deterioration. Pre-operative mean IOP was 23.8 ± 20.3 mmHg (range 6 – 89 mmHg) and decrease to 20.8 ± 6.3 mmHg (range 12 – 34 mmHg) postoperatively (p=0.07). There are no sight-threatening complications found after the surgery. Nevertheless, on the last follow-up, five patients were diagnosed with glaucoma.

A single case report of bilateral iridectomy was published by Chang et al. The optical iridectomy was done in the temporal area of a 4-week-old male infant. Six months after the operation, both eyes showed a good fundus reflex and able to fixate and follow. Postoperative IOP was not stated,
and complication was not found in this study.

DISCUSSION

Worldwide data showed that there is only 2.2 incidence of congenital corneal opacities in 100,000 births affecting visual development, and Peters anomaly took the largest proportion, around 1.5 per 100,000 newborns. There are two types of Peters anomaly, type I characterized by a central corneal opacity with iridocorneal adhesion, and type II is comprised of central corneal opacity with cataract or corneolenticular adhesion. In addition, those with systemic anomalies (cleft lip, short stature, abnormal ears, and mental retardation) and either Peters anomaly type I or type II were included in Peters plus syndrome. Like other congenital anomalies that rarely stand alone, Peters anomaly also associated with several ocular abnormalities, and most commonly were microphthalmia, coloboma, and glaucoma.

The most concerning problem in Peters anomaly is the visual deprivation secondary to central corneal opacity. It will disrupt the normal visual development resulting in sensory deprivation amblyopia. Therefore, it is crucial to treat the central corneal opacity before one-year-old, as the first year of life is the most sensitive period for developing amblyopia. Hitherto, there are several treatments, both pharmacological and surgeries, that have been used to create a clear visual axis in those with central corneal opacities.

Penetrating keratoplasty (PK) was the most commonly performed surgery in a patient with central corneal opacities that affect every layer of the cornea, like in Peters anomaly. In this procedure, all layers of the recipient's cornea will be changed with the donor's cornea. However, performing this technique in an infant's eyes is more challenging than in adult eyes, and pediatric PK requires significant resources in the postoperative period, including biweekly examination under anesthesia and high-dose of topical steroid. Furthermore, in Peters anomaly, this treatment needs to be done before one-year-old, and studies showed that the younger the infant underwent the PK, the higher the incidence of graft failure. Another drawback is the high incidence of glaucoma, occurring in up to 20 - 70% in patients with Peters anomaly, which will negatively affect the success rate of PK. Study by Lucy et al. gave additional information about the lower survival probability for the second and subsequent grafts. Thus, the timing of the surgery is crucial. The delayed timing of PK will increase the graft survival, but at the same time it will also increase the incidence of amblyopia.

Rotating corneal autograft was proposed as an alternative treatment to treat corneal opacities due to its ability to overcome the high incidence of graft rejection in PK. This technique has a lower incidence of graft rejection because the affected cornea of the patients will be changed with the clear peripheral cornea from the patients themselves. However, only a few patients are suitable for this technique, as it needs a minimum diameter of 4 – 5 mm of the unaffected cornea. Thus, this technique cannot be used as an alternative treatment in a patient with Peters anomaly, who usually has a large corneal opacity.

Pharmacological treatment using phenylephrine 1% can be used as an alternative treatment in a small central
leucoma before the definitive treatment, penetrating keratoplasty, took place. It will dilate the pupil, and the clear area surrounding the opacity will serve as a clear visual axis. However, strong adherence is mandatory while using phenylephrine because it has to be used three times a day. Another drawback is its vasopressor effect, which will increase blood pressure. Consequently, the use of phenylephrine as the treatment for central corneal opacity in an infant with Peters anomaly is limited.\textsuperscript{1,2}

Optical iridectomy was proposed as an alternative treatment since the other two options cannot overcome the limitations of PK in Peters anomaly. Optical iridectomy will increase the pupil size providing a larger visual field, which will prevent sensory deprivation amblyopia.\textsuperscript{13} A clear peripheral corneal is needed to perform the optical iridectomy, and preferably not in the superior quadrant, which can be covered by the upper eyelid.\textsuperscript{9,14} This technique has several advantages, as it can be performed by a general ophthalmologist in the area where corneal transplantation facility is not available. Postoperative care is much easier than PK without the risk of having graft rejection or secondary glaucoma due to high-dose topical steroids.\textsuperscript{15} All studies showed that optical iridectomy is a safe procedure that can even be done in a one-week-old infant.\textsuperscript{1} In addition, three studies that were included in this review showed a promising result of visual improvement, and two of those also showed a good intraocular pressure postoperatively.\textsuperscript{2,9,16}

Limitations of our study is the small amount of sample. However, this studies have included all studies that report the outcome of optical iridectomy in Peters anomaly. Further study regarding the long-term outcome of visual acuity and adverse effect is still needed.

**CONCLUSION**

A clear visual axis, preferably before one-year-old, is mandatory in patient with Peters anomaly to prevent the occurrence of sensory deprivation amblyopia. Optical iridectomy can be used as a simple and safe alternative treatment to achieve clear visual axis in those who have clear peripheral cornea.
Table 1. Characteristics of the studies

<table>
<thead>
<tr>
<th>Author (year)</th>
<th>Number of patients</th>
<th>Age</th>
<th>Study design</th>
<th>Markers of improvement</th>
<th>Results</th>
<th>Post Operative IOP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sprieger O, et al. (2017)</td>
<td>22</td>
<td>1 week – 7.7 years</td>
<td>Retrospective case series</td>
<td>Red reflex Visual acuity</td>
<td>Improvement</td>
<td>Decrease</td>
</tr>
<tr>
<td>Chang TC, et al. (2018)</td>
<td>1</td>
<td>4 weeks</td>
<td>Case reports</td>
<td>Red reflex Fix and follow test</td>
<td>Improvement</td>
<td>Not stated</td>
</tr>
</tbody>
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REFERENCES