Table of contents

Original articles

50 Evaluation of the filtering bleb and its relation to an intrascleral aqueous drainage route after trabeculectomy using ultrasound biomicroscopy
Ay Ser A. El-Hameed Fayed, Tarek T. Soliman

54 Primary versus secondary intraocular lens implantation in the management of congenital cataract
Thanaa H. Mohamed, Rania G. Eldin Zaki, Mohamed H. Hashem

60 LASIK in selected patients with posterior polymorphous corneal dystrophy
Shereef Abdelwahab, Maha Elfayoumi

65 Photorefractive keratectomy with corneal collagen cross-linking 1 year after intrastromal corneal ring segment implantation for the treatment of keratoconus
Maha Elfayoumi, Shereef Abdelwahab

71 Evaluation of a new multifunctional vitreoretinal twister for use in advanced proliferative diabetic retinopathy
Dikran G. Hovaghimian, Saleh Sherif Abd, Tarek Saleh, Mohamed Tarek El-Naggar

85 Surgical management of primary congenital glaucoma in egypt
Nader H.L. Bayoumi

93 Comparative study between big-bubble technique and manual dissection in deep anterior lamellar keratoplasty
Amr S. Abdel Hakeem, Tarek A. Katamish, Dalia H. Khall, Hoda A. Aisaedlin
LASIK in selected patients with posterior polymorphous corneal dystrophy
Shereef Abdelwahab, Maha Elfayoumi

Department of Ophthalmology, Benha University, Benha, Egypt

Correspondence to Shereef Abdelwahab, BSc, MSc, PhD Ophthalmology, 8 Nakhla El Motey Street, Heliopolis, Cairo, Egypt.
Tel: +20 101 456 8919; fax: +20 279 415 38
E-mail: sherif.abdelwahab@fmed.bu.edu.eg

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Purpose
The aim of the present study was to evaluate the safety of LASIK surgery in patients with posterior polymorphous corneal dystrophy.

Patients and methods
A total of 25 eyes of 13 patients with posterior polymorphous dystrophy with various refractive errors were selected for LASIK treatment and were followed up for 2 years after surgery.

Results
All eyes included in the study showed an improvement in unaided visual acuity with no postoperative complications.

Conclusion
LASIK can be a safe procedure in patients with posterior polymorphous dystrophy and proper selection criteria.

Keywords: corneal, dystrophy, LASIK, posterior polymorphous, posterior polymorphous corneal dystrophy

Introduction
Posterior polymorphous corneal dystrophy (PPCD) is a dominantly inherited disorder of the corneal endothelium [1] affecting primarily the corneal endothelium and Descemet's membrane [2]. The clinical expression of this autosomal dominant disorder varies considerably, even within the same family: one family member may be asymptomatic with a single endothelial lesion, whereas a sibling may have corneal decompensation and advanced glaucoma [3].

PPCD can be seen on slit-lamp examination as one of three patterns: vesicle lesions, band lesions, and diffuse opacities. The most common is the vesicular lesion, which is present in almost all patients with PPCD. The vesicular lesion appears as a transparent cyst and can be found anywhere on the posterior cornea and may appear as isolated lesions or in lines, clusters, or confluent groups [4]. By specular microscopy, the vesicular lesions range in diameter from 0.10 to 1.0 mm [5].

Band lesions are typically horizontal, have parallel scalloped edges, and do not taper toward the ends and can be found on any area of the posterior cornea. Specular microscopy of the band lesions shows shallow trenches and ridges formed from a large number of confluent vesicles [5].

Diffuse PPCD appears as small, gray–white lesions or larger geographic lesions anywhere on Descemet's membrane. With direct illumination, there is a haze in the posterior stroma adjacent to the lesions. Under retroillumination, the opacities have a peau d'orange texture [6].

Slit-lamp findings reveal corneal edema in the more advanced cases, and calcific and lipid-degenerative changes in severe cases. Iridocorneal adhesions, if present, can range in severity from fine or broad-based adhesions seen only on gonioscopy to large iridocorneal adhesions often associated with a glass-like membrane that are seen easily by slit-lamp examination. All patients with broad-based iridocorneal adhesions have an elevated intraocular pressure [7].

PPCD is a bilateral disease, and is usually asymmetric, and most cases are asymptomatic and stable and diagnosed on incidental slit-lamp examination. This dystrophy usually occurs in the second or the third decade of life and rarely appears earlier.

The term posterior corneal vesicle syndrome has been coined for the presence of unilateral vesicular or band lesions similar to PPCD in patients, but which is not found in other family members [8].
Patients with PPCD seeking refractive surgery must be examined carefully to determine whether he or she is a good candidate for surgery. The major concern in evaluating these patients is whether the patient has active corneal edema during the preoperative evaluation. An endothelial cell count is taken, and if this cell count is significantly diminished, then surgery would mostly be a contraindicated procedure. The corneal thickness is an important parameter to help decide whether a patient can undergo refractive surgery; pachymetry and a Pentacam measurement could determine the corneal thickness and whether there are any local variations in the thickness that could be undetectable at the slit lamp. If the pachymetry is normal, the patient has good corrected vision, with a clear cornea and within normal endothelial cell density (ECD), and a normal percent of hexagonal endothelial cells, it is most likely that he/she can undergo LASIK.

Patients and methods

Patients in this study were treated according to an approved protocol. All patients signed an informed consent form before each procedure.

A total of 25 eyes of 13 myopic and two hyperopic patients were included in this study. All patients were diagnosed to have PPCD at the slit lamp incidentally on seeking refractive surgery. Patients included in the study had no clinically detected corneal edema and normal open angles on gonioscopy, with normal intraocular pressure and optic nerve heads and no clinical sings of glaucoma. Both vesicular lesions (18 eyes) and band lesions (seven eyes) were included in the study and none of the diffuse lesions were included. All lesions were located in the peripheral or the paracentral cornea outside the central 6 mm. All patients were subjected to routine preoperative LASIK investigations including, aided and unaided visual acuity assessment, refraction both with and without cycloplegia, slit-lamp examination, gonioscopy, intraocular pressure measurement using the Goldman Applanation Tonometer; Haag-Streit, USA, and corneal tomography using the Allegro Oculyzer; Wavelight AG [ALCON LABORATORIES], Erlangen, Germany, with the Belin-Ambrosio enhanced ectasia display software. Patients with a central corneal thickness between 500 and 620 μm were only included in the study. Specular microscopy for ECD and the percent of pleomorphism using the Nidek CEM-530 specular microscope was carried out for all patients; a minimum ECD of 2800/mm² was set as a limit for inclusion in the study.

The surgical procedure

All patients had there LASIK procedure performed using the WaveLight Allegretto Wave Eye-Q (Wavelight AG [ALCON LABORATORIES], Erlangen, Germany) 400 Hz platform. After sterilization and draping, topical oxybuprocaine 0.4% was applied. LASIK flaps were created using the MoriaM2 microkeratome using the disposable single-use 90 blade. All the cases were uncomplicated straightforward LASIK procedures. Postoperatively, topical antibiotic drops, moxifloxacin hydrochloride 0.5% (Vigamox; Alcon a Novartis company), and steroid drops, prednisolone acetate 1% (Pred Forte; Predforte eye drops, Allergan, Inc, Irvine, CA92612, USA), were prescribed every 6 h for 1 week. Artificial tears were prescribed every 4 h for 1-month postoperatively and the dose was then adjusted or stopped accordingly.

Postoperative follow-up

All patients were seen on the first postoperative day, after 1 week, after 1 month, and then every 6 months until the end of the scheduled 2-year follow-up period. In each follow-up, unaided visual acuity was recorded, and a slit-lamp examination was performed. Specular microscopy recording ECD and the percent of pleomorphism were determined at 1 week, 6 months, and in the last follow-up visit at the end of the 2-year follow-up.

Results

Preoperative and postoperative results were compared using the data analysis tool pack in Microsoft Excel using the paired two-sample t-test.

A total of 25 eyes of 13 patients [eight (61.5%) female and five (38.5%) male] were included in this study; the mean age of the male patients was 28.5±1.44 years and that of the female patients was 30.1±2.72 years. About 11 patients were myopic and two were hyperopic. The mean myopic error was −3.5±0.98 (range: −0.0 to −0.0); five of these patients had a mean myopic astigmatic error of −1.7±0.65 D cylinder (range: −0.5 to −0.0 D cylinder). The mean hyperopic error was +4.0 (range: +3.0 to +5.0). All patients came seeking refractive surgical correction. PPCD was diagnosed on slit-lamp examination, and none of the participants had any symptoms related to PPCD. About 18 (72%) eyes were of the vesicular lesion type and eight (28%) eyes were of the band lesion type. The mean preoperative uncorrected visual acuity (UCVA) was 20/400 (range: 20/500–20/80). The mean preoperative central corneal thickness was 536±7.7 μm (range: 501–610 μm) and the mean ECD was 3019 cells/mm² (range: 2880–3320 cells/mm²). The mean preoperative percent of endothelial
hexagonal cells was 73±3% (range: 70–76%). In the first postoperative visit, the mean UCVA was 20/25 (range: 20/30–20/15), the cornea was clear in all cases, and no corneal edema was noticed on slit-lamp examination. In the final follow-up visit, all eyes had a mean UCVA of 20/20 (range: 20/25–20/15) and a mean ECD of 2979 cells/mm² (range: 2860–3312 cells/mm²) with a mean reduction of 1.3% (P>0.5), which was highly insignificant. The mean percent of endothelial hexagonal cells was 72.7±2% (range: 70.7–74.7%; P>0.5), which was highly insignificant (Figs 1–3).

**Discussion**

The refractive surgeon is insignificant faced with patients seeking refractive surgical correction, some of whom happen to have a corneal pathology that could be an absolute surgical contraindication. Other pathologies such as PPCD, an autosomal dominant disorder affecting the corneal endothelium, and Descemet’s membrane, are not believed to be an absolute contraindication. The question about whether to proceed with the surgery in such cases or not is a matter of debate. As the main pathology is in the Descemet’s membrane and the corneal endothelium, which play a vital role in maintaining the corneal clarity, any damage to either of them could affect the optical corneal clarity and hinder the result of any refractive procedure. Risks to the endothelium caused by the excimer laser include mechanical trauma from shockwaves, local oxidative changes, and thermal effects from ultraviolet radiation [9,10]. Marshall et al. [11] mentioned that 193-nm excimer laser treatments to the superficial cornea do not damage the endothelium. Kim et al. [12] demonstrated that a residual stromal bed of ∼200 μm prevents endothelial cell damage. This 200-μm threshold is generally not reached by laser treatments today because surgeons calculate and measure the residual stromal bed thickness, and a higher threshold of at least 300 μm is recommended to avoid corneal ectasia [13]. Overall, the evidence suggests that laser refractive surgery does not affect the corneal endothelium adversely [14].

Dastjerdi and Sugar [15] reported that laser in-situ keratomileusis can result in corneal endothelial decompensation in patients with Fuchs endothelial dystrophy. They attribute the damage to the combination of excimer laser ablation and increased intraocular pressure (IOP) associated with the LASIK procedure, which exerts a cumulative traumatic force on the corneal endothelium resulting in edema.

Huerva and Mateo [16] report rejecting refractive surgery in a 50-year-old female patient who presented for refractive surgery consultation, but was found on examination to have PPCD. A low endothelial cell count may be a contraindication for
a LASIK procedure in cases of posterior polymorphous dystrophy because of the possible risk of corneal decompensation. The authors provide a review of reported endothelial changes with LASIK and photorefractive keratectomy and recommend against performing either corneal refractive procedure in patients with PPCD.

Moshirfar et al. [2] report the result of their study on two patients with PPCD for whom they performed LASIK refractive correction. One-year postoperatively, all eyes had a UCVA of 20/25 or better and a corrected visual acuity of 20/20 or better. There was a mean endothelial cell loss of 2.3% at 1 year, which was greater than the expected 0.5% annual loss, but no corneal decompensation or other complications were reported. The authors conclude that ‘performing LASIK in patients with PPCD is likely safe in the short term, when performed on patients who have the vesicular type of PPCD, no central lesions, and near-normal ECD’. Bower et al. [17] studied the safety and the efficacy of photorefractive keratectomy in 14 patients with PPCD with vesicular and band subtypes. They concluded that ‘Photorefractive keratectomy in PPCD patients with vesicular and band subtypes resulted in excellent visual outcomes and a low incidence of adverse effects’.

In the present study, we studied the safety of LASIK in selected cases with PPCD; all our patients had a refractive error (11 myopes and two hyperopes) associated with either vesicular or band-type PPCD that was not located in the central cornea. A minimum ECD of 2800 cells/mm² was allowed. The cell density alone is not the most sensitive measure of endothelial health, as the endothelium functions even at low ECDs (under 500 cells/mm²) [17]. Pleomorphism, a variation in the cell shape, as represented by the percentage of hexagonal cells, is a more sensitive measure of the endothelium under stress. In the normal healthy cornea, about 70–80% of the endothelial cells are hexagonal. The mean percentage of hexagonal endothelial cells in the present study was 73%.

The final outcome of our study was in favor of LASIK in selected patients with PPCD, in contrast to the study of Dastjerdi and Sugar [15], which was conducted on a 47-year-old woman with Fuchs endothelial dystrophy and cornea guttata. Huerva and Mateo [16] also did not recommend LASIK for a patient with PPCD. The first study dealt with a patient with Fuch’s endothelial dystrophy with cornea guttata clinically diagnosed before surgery. The second study denied LASIK surgery to a 50-year-old female patient with PPCD attributing this to a detected lower than normal ECD.

Our results are close to the results of Moshirfar et al. [2] who found LASIK in patients with PPCD is likely safe in the short term, when performed on patients who have the vesicular type of PPCD, no central lesions, and near-normal ECD. Their study was close to our study regarding the selection criteria, yet it was conducted on only four eyes with the vesicular type of PPCD, and their cases were followed up for 1-year postoperatively. This study included 25 eyes of 13 patients with both vesicular-type and band-type PPCD, and our follow-up period was 2 years.

No significant changes in ECD or endothelial pleomorphism as a result of LASIK surgery were found in this study. Three hyperopic eyes were included in the study and no complications were recorded; all previous studies conducted on patients with PPCD were performed on myopic patients. The main difference between LASIK in myopic and hyperopic patients is the ablation zone being larger and more toward the periphery in hyperopic LASIK correction. The number of hyperopic eyes included in this study is however quite few, and further studies evaluating the safety of LASIK in patients with PPCD and hyperopia are recommended. There was a statistically insignificant reduction of 1.3% in ECD at the end of the 2-year follow-up, which is within the normal age-related cell loss rate of 0.3–0.6% per year [14].

These results are applicable to carefully selected cases of PPCD, such as in cases with an ECD of 2800 cells/mm² or higher and in cases without glaucoma, iris abnormalities, or corneal decompensation, with the lesion out of the central cornea and good endothelial cell hexagonal pleomorphism percent.

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Conflicts of interest
There are no conflicts of interest.

References