

# High ablation depth phototherapeutic keratectomy in an advanced case of Reis-Bucklers' corneal dystrophy

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## ARTICLE INFO

### Keywords:

Reis-Bucklers' corneal dystrophy  
Phototherapeutic keratectomy  
Recurrence  
High ablation depth

## ABSTRACT

**Purpose:** Phototherapeutic keratectomy (PTK) plays an important role in the management of Reis-Bucklers' corneal dystrophy. The usage of PTK for the treatment of anterior corneal dystrophies is limited by the depth of ablation. This case shows the usage of high ablation depth PTK as one of the management strategies to visually rehabilitate a patient of advanced Reis-Bucklers' corneal dystrophy.

**Observations:** We present a case of 32-year-old lady who was diagnosed with Reis-Bucklers' dystrophy in both eyes. The patient had advanced form of the disease and history of recurrent corneal erosions. For visual rehabilitation and symptomatic relief, phototherapeutic keratectomy was planned in both eyes. In view of the advanced nature of the disease, the ablation depth had to be high to get an acceptably clear central cornea.

**Conclusions and Importance:** PTK with high ablation depth is possible in advanced Reis-Bucklers' corneal dystrophy unlike other types of anterior corneal dystrophies.

## 1. Introduction

Corneal dystrophies of Bowman's membrane, most commonly Reis-Bucklers' dystrophy, involve the superficial stroma and cause poor vision from corneal irregularity and opacification. Reis-Bucklers' dystrophy is bilateral autosomal dominant corneal dystrophy caused by a specific mutation in the TGFBI gene (5q31).<sup>1</sup> It generally presents early in life with recurrent corneal erosions leading to pain and photophobia.<sup>2</sup> The initial clinical picture is that of discrete anterior corneal opacities at the level of the Bowman's membrane which subsequently become more confluent.<sup>3</sup> Reis-Bucklers' dystrophy's nomenclature has been a bit confusing as it has had the following names: granular corneal dystrophy [GCD] type III, superficial variant of GCD, corneal dystrophy of Bowman layer type I.<sup>4</sup> Recently, the International Committee for Classification of Corneal Dystrophies (IC3D) has categorized these dystrophies as "epithelial-stromal TGFBI dystrophies".<sup>5</sup> Phototherapeutic keratectomy (PTK) is performed to clear central corneal opacities and prevent recurrent corneal erosions.<sup>6</sup> PTK is accomplished using a 193-nm excimer laser to ablate the least amount of tissue and clear maximum amount of central cornea. PTK causes remarkable improvement in vision and provides symptomatic relief to the patient. The recurrence rate after

the treatment is quite high.<sup>7</sup> Still PTK is a less invasive surgical modality as compared to corneal transplantation and it can be repeated later if recurrence occurs. The application of Mitomycin C (MMC) has been shown to reduce the chances and slow the recurrence of the dystrophy after PTK.<sup>8,9</sup> This case highlights the usage of PTK with high ablation depth and adjuvant MMC use as a relatively less invasive treatment modality for an advanced case of Reis-Bucklers' corneal dystrophy.

## 2. Case report

A 32-year-old lady presented with Reis-Bucklers' corneal dystrophy in both eyes. The patient had history of progressive diminution of vision since the age of 6 years and attacks of recurrent corneal erosions. The uncorrected visual acuity (UCVA) was 20/800 in both eyes which was not improving with refraction or pinhole.

Slit lamp examination of both eyes showed dense diffuse grayish deep epithelial opacities involving the visual axis in a fishnet pattern [Fig. 1 (a) and (c)]. Anterior segment optical coherence tomography (AS-OCT) imaging of both eyes cornea showed a thick band of hyper-reflective material in the epithelial and subepithelial area which was 158  $\mu\text{m}$  thick in right eye and 144  $\mu\text{m}$  thick in left eye including the

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<https://doi.org/10.1016/j.ajoc.2022.101299>

Received 20 October 2020; Received in revised form 7 June 2021; Accepted 17 January 2022

Available online 20 January 2022

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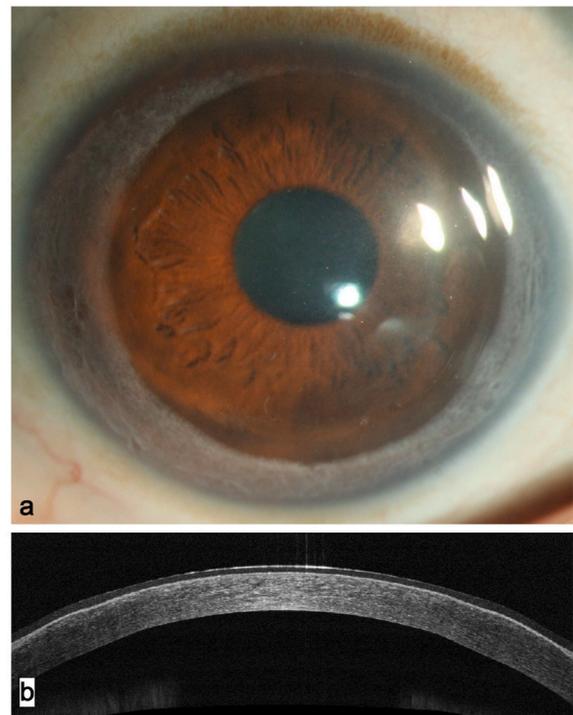
epithelium, bowman's membrane, and anterior stroma [Fig. 1(b) and (d)]. Rigid Gas Permeable (RGP) contact lens (CL) trial was done for both eyes to evaluate the visual potential. Even scleral CL (Prosthetic Replacement of Ocular Surface Ecosystem, PROSE) trial was done which improved the vision further with more comfort. But the patient was not willing for usage of CL.

PTK was planned for the visual rehabilitation and symptomatic comfort of the patient, right eye followed by left eye 2 weeks later. Transepithelial PTK using the Wavelight EX500 excimer laser platform (Alcon Inc., Fort Worth, Texas, USA) was performed. The optic zone was set at 7mm and transition zone at 1.25mm. The cumulative ablation depth for the right eye was 150  $\mu\text{m}$  and for the left eye was 140  $\mu\text{m}$ . Subsequently, a sponge, 7.0 mm in diameter, soaked with 0.02% MMC (0.2 mg/mL, diluted in balanced salt solution), was applied over the ablated surface for a duration of 90 seconds. Thorough corneal surface wash was given with 30ml balanced salt solution, and a bandage contact lens (BCL) was placed. Slit lamp examination at the end of the procedure showed clear central cornea in both eyes with BCL in situ (Fig. 2).

The post-operative recovery was uneventful. The BCL was removed after 1 week. The uncorrected distance visual acuity (UDVA) at 1 month in both eyes were 20/80 which was improving to 20/30 with refraction of +1.25D sphere in right eye and +1D in left eye. The patient was also extremely comfortable. The patient was followed up regularly thereafter every 6 months to keep a watch on recurrence. At 2-year follow-up, there was evidence of very early recurrence in both eyes, but the patient was visually and symptomatically comfortable (Fig. 3). The BCVA in both eyes was 20/40 with manifest refraction of +1.75D sphere in both eyes. The patient has been under regular follow-up since last 4 years. There was early asymptomatic recurrence in both eyes. The endothelial cell density and morphology was also normal in both eyes. The BCVA in both eyes was 20/50 with refraction of +2.25D sphere in both eyes at last follow-up.

### 3. Discussion and conclusions

The utility of PTK for treatment of Reis-Bucklers' corneal dystrophy has been well documented in literature.<sup>6,10</sup> Miller et al.<sup>9</sup> reported the use of excimer laser PTK for treatment of Reis-Bucklers' dystrophy with a single intraoperative application of adjunctive topical 0.02% MMC.

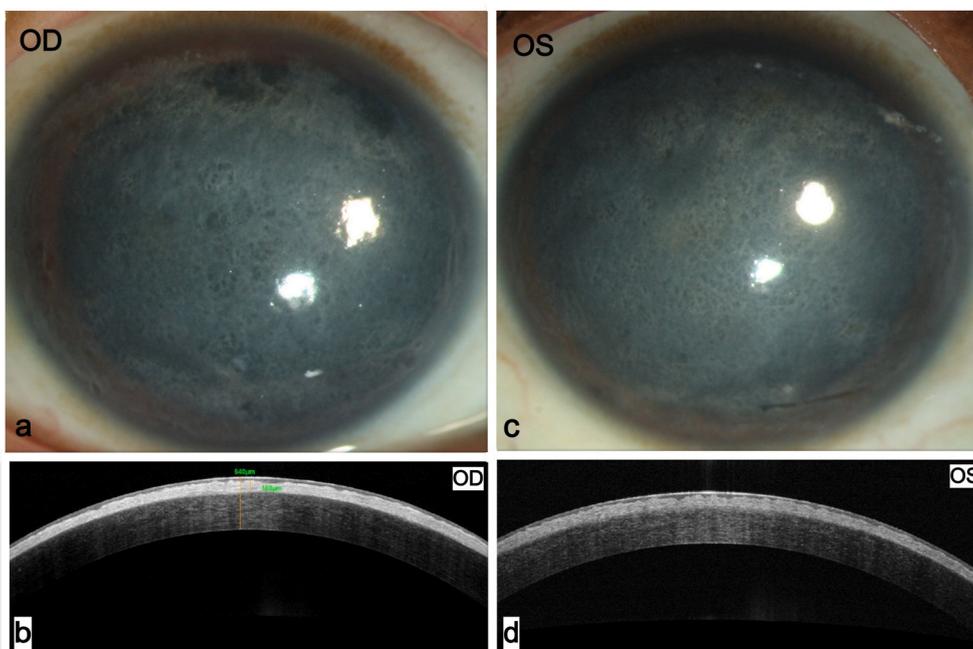


**Fig. 2.** Title: Clinical picture after PTK.

Legend: (a) Slit lamp image of the right eye cornea after PTK showing central area free of deposits and BCL in situ. (b) AS-OCT of the right eye cornea showing absence of deposits in the central 7mm of cornea after PTK.

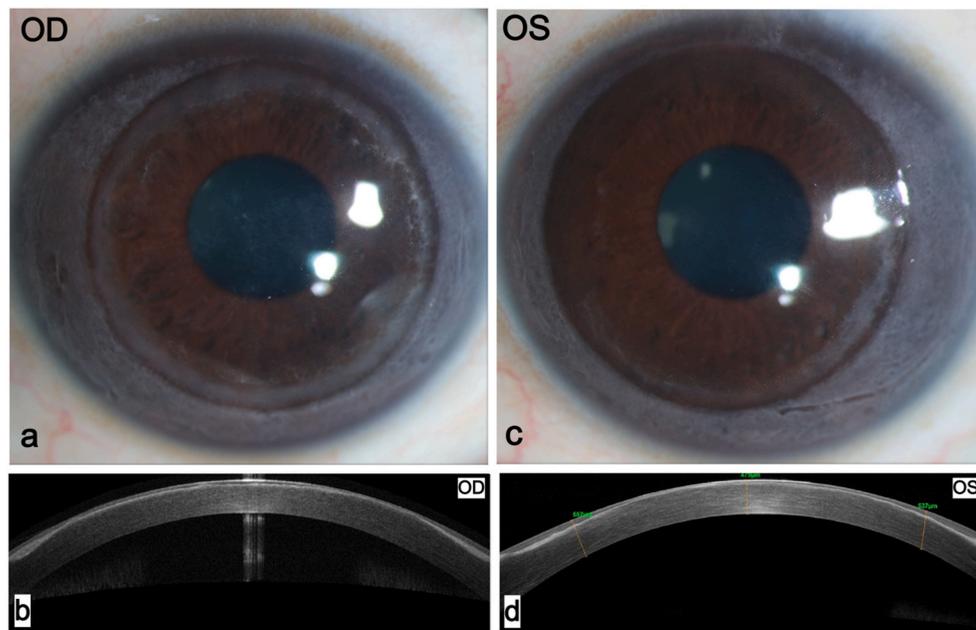
They postulated that use of topical MMC in conjunction with PTK may prevent the recurrence of Reis-Bucklers' dystrophy. McDonnell and Seiler<sup>5</sup> described two cases of Reis Bucklers' dystrophy treated by PTK without masking fluid and found satisfactory results.

There are no set guidelines as to maximum depth of ablation in PTK. Most of the PTK procedures performed involve corneal stromal ablation of between 40 to 100  $\mu\text{m}$ .<sup>7</sup> Keeping in mind the risk of ectasia, it has been advised to have the residual stromal bed thickness of at least 250



**Fig. 1.** Title: Clinical picture at presentation.

Legend: (a) Diffuse illumination slit lamp photograph of the right eye cornea showing greyish reticular opacification extending from limbus to limbus. (b) AS-OCT of the right eye cornea showing hyperreflective membrane like subepithelial deposits with clear underlying stroma. (c) Diffuse illumination image of left eye cornea showing the fishnet pattern opacification. (d) AS-OCT of the left eye cornea showing the homogenous subepithelial band like deposits.



**Fig. 3.** Title: Early recurrence after 2 years.

Legend: (a) Diffuse illumination slit lamp image of right eye cornea showing early recurrence mainly in the peripheral zone of ablation. (b) AS-OCT showing very thin hyperreflective subepithelial deposit. (c) Early peripheral recurrence in left eye. (d) AS-OCT showing the peripheral deposits in the left eye cornea.

$\mu\text{m}$  after the procedure. Deep stromal ablation is associated with increased risk of induced hyperopia after the procedure. An ablation depth of less than  $100 \mu\text{m}$  is less likely to induce significant hyperopia.<sup>11</sup> Higher ablation depth and delayed epithelial healing predispose to post-operative haze formation after PTK.<sup>12</sup>

The overall depth of the central corneal opacity due to Reis-Bucklers' dystrophy in our case was about  $150 \mu\text{m}$  in both eyes. The patient had not sought any surgical intervention for her eye condition since childhood. As a result of this, the corneal subepithelial deposits had become quite thick over a period. The residual clear stroma beneath the opacity was measured to be around  $480 \mu\text{m}$  on AS-OCT. Hence, we decided to plan PTK for this patient, even though it involved high ablation depth. Lamellar keratoplasty is also one of the treatment options for advanced cases of Reis-Buckler's corneal dystrophy.<sup>13</sup> With the advent of femtosecond laser usage for anterior lamellar keratoplasty, the surgery has become more predictable and technically less challenging. Shousha et al.<sup>14</sup> have shown that femtosecond laser-assisted sutureless anterior lamellar keratoplasty (FALK) is a favorable option for anterior corneal pathologies with rapid visual rehabilitation and minor induced astigmatism.

The anterior surface of the cornea in our patient was smooth as seen on AS-OCT which itself acted as a masking agent. Also, there is a risk of increase in the surface irregularity with manual removal of epithelium.<sup>15</sup> So, we used PTK for epithelial removal as opposed to manual removal. Transepithelial ablation is advisable in anterior corneal stromal dystrophies.<sup>16</sup> This also helped to avoid the use of masking agent which can make adequate ablation depth determination difficult. To reduce the chances of post-operative scarring, we used intra-operative MMC after the ablation. There is evidence that usage of topical MMC application after PTK for Reis-Buckler's dystrophy reduces the chances of recurrence.<sup>8,9</sup> The duration of 90 seconds was determined since the total ablation depth was around  $150 \mu\text{m}$  which roughly corresponds to 10D of refractive error correction in refractive ablation. We follow the rule of 10 seconds MMC exposure per diopter correction of refractive ablation. In all cases of PTK, we use 90 seconds of MMC exposure irrespective of the ablation depth.

In advanced cases of Reis-Bucklers' corneal dystrophy, the deposits replacing the basal epithelial cells and the Bowman's membrane are

accumulated over time. This provides additional thickness to the anterior cornea with relatively uninvolved mid to deep stroma. Hence in these cases, PTK with higher ablation depth can be planned safely as it will ablate the added deposits and not deep into the stroma. The usage of MMC seems to a good idea to reduce the chances of haze and provides added advantage of reducing recurrence in Reis-Bucklers' corneal dystrophy.

To the best of our knowledge, this is the first case report in literature highlighting the use of high depth PTK for an advanced case of Reis-Bucklers' corneal dystrophy. To have a good visual outcome in these cases, high ablation depth can be attempted. Also, the procedure is repeatable and helps delay more invasive corneal transplantation.

#### Patient consent

Written informed consent to publication (including images, personal and clinical details of the participants) has been obtained from the patient.

#### Funding

Hyderabad Eye Research Foundation, India.

#### Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

#### Declaration of competing interest

None.

#### Acknowledgements

No additional acknowledgements.

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