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Scleral contact lenses in the management of pellucid marginal degeneration

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ABSTRACT

Purpose: To assess visual improvement with scleral lenses (PROSE, prosthetic replacement of the ocular surface ecosystem, Boston Foundation for Sightight, USA) in patients with Pellucid marginal degeneration (PMD).
Methods: This is a single-center, retrospective case-series involving patients with clinical PMD who underwent scleral lens-PROSE trial for improvement of visual acuity, from January 2009 to December 2012 at a tertiary center in India. Scleral lenses with different front surface eccentricities (FSE) were tried for improvement of visual acuity. Snellen visual acuity before and with PROSE wear was noted. Complications with PROSE wear to follow-up were noted.
Results: PROSE was dispensed to 12 patients (20 eyes) out of 19 patients having PMD. Location of PMD was inferior in fourteen and superior in two eyes. Four eyes had co-existing keratoconus. Nine were males and three were females. The indications for scleral lens were lens popping-out, failure of piggy-back contact lens and RGP failure. LogMAR Visual acuity improved significantly from 0.45 ± 0.31 pre-PROSE to 0.05 ± 0.08 post-PROSE ($p = 0.0001$). The FSE ordered was 0.6 in 17 eyes, 0.3 in one eye and 0.8 in two eyes. Three patients had hydrops over follow-up; two patients underwent keratoplasty and one was managed conservatively with steroids and hyperosmotic agents. Seven patients did not order PROSE: reasons were - no perceived improvement in visual acuity ($n = 2$), wanted to decide ($n = 2$), continued glasses ($n = 1$) and continued RGP contact lens ($n = 1$). One patient had difficulty with self lens insertion.
Conclusion: PROSE improves visual acuity in PMD; three patients developed hydrops over follow-up.

1. Introduction

Pellucid marginal degeneration (PMD) is a bilateral non-inflammatory ectatic disorder of the cornea characterized by corneal ectasia above a peripheral band of thinning of cornea, usually inferiorly, from 4.00 to 8.00 O' clock, 1–2 mm away from the limbus [1]. PMD usually presents between 20 and 40 years of life [1–4]. It is usually bilateral but unilateral cases are reported [5,6]. The etiology of PMD is unknown. Histopathological studies have reported irregular Bowman's membrane or breaks in Bowman's layer, stromal thinning and the absence of inflammation. The complications include hydrops and corneal perforation [7].

The ectasia results in high against the rule astigmatism resulting in progressive diminution of both, uncorrected as well as best-corrected visual acuity (BCVA) in these patients [8]. The correction of the irregular astigmatism remains a challenge. Spectacles may play a role in the very early stage of the disease [6]. However, as the condition progresses, contact lenses are imperative for visual improvement. In the early stages of PMD, improvement in visual acuity is possible with soft toric lenses. However, with progression of disease, customized soft toric lenses, rigid gas permeable (RGP) contact lenses, bitoric lenses, reverse geometry lenses, piggyback contact lenses (PBCL), hybrid lenses and scleral lenses may be useful [7,9–14]. Surgical modalities include penetrating keratoplasty (PK), which is done as last resort, lamellar or crescentic lamellar keratoplasty, wedge excision, banana graft first followed by optical PK, tuck-in lamellar keratoplasty, phakic IOLs and intracorneal ring segments

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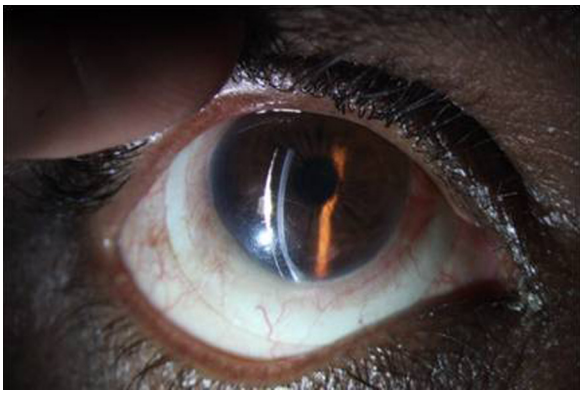

Fig. 1. PROSE in eye of a patient with inferior pellucid marginal degeneration.

implantation [2,7,15]. Most often, surgeries are indicated if a patient is contact lens intolerant, there is no improvement in vision with contact lenses, failure to obtain an acceptable fitting or presence of scars [7]. Moshirfar et al. has described the guidelines for the management of PMD in a nomogram under different criteria such as cornea- independent, structural, full thickness and partial thickness interventions [7].

Scleral lenses are large diameter lenses, which vault the cornea and rest on the sclera [16]. There is a fluid reservoir between the lens and the cornea [16]. Scleral lenses are indicated when visual acuity does not improve with the traditional contact lenses or fitting is not possible with various available contact lens options [2,3,17]. PROSE (prosthetic replacement of the ocular surface ecosystem, Boston Foundation for Sight, USA) is custom designed by using a proprietary computer-assisted design and manufacture (CAD/CAM) software, linked to a computerized lathe machine, which uses spline technology to create a junction-less posterior surface [18].

Various studies have reported improvement in visual acuity with PROSE in patients having corneal ectasia such as keratoconus and PMD [10,11]. PMD is mostly grouped as irregular astigmatism other than keratoconus. [14,18–21]. However, to the best of our knowledge, there are no studies citing the results of PROSE in patients with PMD. The aim of this retrospective study is to assess improvement in visual acuity with scleral lenses i.e. PROSE in patients having PMD.

2. Materials and methods

This was a retrospective study conducted for the patients seen in the scleral lens clinic from January 2009 to December 2012 at a tertiary eye care center in India. Institute Review Board of L V Prasad Eye Institute, Hyderabad, India approved the study.

2.1. Subjects

Of the total 941 patients seen in the scleral lens clinic, 19 patients having PMD underwent PROSE trial for improvement of visual acuity. All patients were seen by the Cornea Specialists and were referred from Bausch & Lomb Contact Lens Clinic at the L V Prasad Eye Institute, Hyderabad, India. PMD was clinically diagnosed, based on the peripheral band of thinning–either superior or inferior, with a clear area in between the thinned area and the limbus and presence of ectasia inferior or superior to the area of thinning, respectively. The corneal topography was done when possible with Orbscan IIz (Bausch & Lomb, Rochester, NY).

2.2. Fitting of PROSE

Fitting of PROSE has been already described in literature [22]. Initially we used the scleral lenses of 18 mm diameter, but later on, the lenses of 18.5 mm diameter were used. Scleral lenses with larger diameter (greater than 18.0 mm) are preferred in most cases as they avoid the development of excessive suction between the lens and the ocular surface [23,24]. The first lens was selected based on the ease of insertion. The large diameter of the lens may intimidate the patients. PROSE was inserted in the eye after filling with normal saline. This was followed by assessing patient's comfort. Slit lamp biomicroscopy was done to assess the fitting. Assessment included a proper alignment of the haptic or the scleral portion of the lens to the sclera, corneal clearance and an optimal edge alignment without causing any impingement on the conjunctiva, or edge lift of scleral lens. The fitting procedure included examining the patient immediately, after one hour of the lens wear and after 4 h of the lens wear. Visual acuity was assessed after 4 h. PROSE with different front surface eccentricity values (FSE) were tried to improve vision.

The over refraction (with loose lens in trial frame) was done and added to the PROSE power before ordering it. Presence or absence of conjunctival staining was noted immediately after PROSE removal from the eye. Tenderness was checked through lids if the staining of the conjunctiva was positive. This aided us in designing a toric haptic for PROSE as the presence of conjunctival staining in one area indicated that the haptic of the scleral lens needed to be flattened in this area and toricity needed in the opposite meridian. Care was taken to note that there was no suction felt when the fitter removed the PROSE after four hours of wear. The final fitting orders were made based on—the patient's comfort, the alignment of the haptic or scleral portion of the lens to the scleral, an adequate corneal clearance, the amount of suction felt by the fitter and improvement in vision.

Demographics in the form of age, gender, the location of PMD, BCVA with habitual correction and with PROSE wear at the last follow-up were noted. Snellen visual acuity was converted to Log MAR visual acuity for statistical analysis, using Student's paired *t* test. A *p* value of 0.05 was considered statistically significant. Statistical analysis was done using Microsoft Excel version 8.1.

3. Results

Twelve (20 eyes) out of 19 patients (28 eyes) received the PROSE lenses (Fig. 1). The mean age of these patients was 43 years (range 29–64 years). There were nine males and three females. Eight patients had bilateral PMD and four had unilateral PMD. The

Table 1
Habitual correction prior to PROSE wear.

Habitual correction	Percentage
Nil correction	20%
Spectacles	10%
RGP contact lenses	45%
Soft contact lenses	20%
Piggy-back contact lenses	5%

Table 2
Reasons for not ordering the PROSE after trial.

Reasons	Number of patients
No perceived improvement in visual acuity	$n=2$
Wanted to decide	$n=2$
Continued glasses	$n=1$
Continued RGP contact lens wear	$n=1$
Difficulty in lens insertion	$n=1$