Use of Scleral Lenses in the Visual Rehabilitation of Keratoconus: A Case Series

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Abstract

Introduction: Scleral lenses also known as scleral contact lenses are rigid gas permeable contact lenses that rest entirely on the sclera without touching the cornea. This unique feature of scleral lenses makes it ideal in the management of complex corneal and ocular surface irregularity.

Purpose: To report the indications, use and fitting of Europa scleral lenses for the visual rehabilitation of two patients with Keratoconus in a specialty contact lens practice in Abuja, Nigeria.

Case reports: We report the cases of two female patients of Nigerian descent diagnosed with Keratoconus who were managed previously with other modalities of contact lenses but presented with unsatisfactory vision and discomfort with their habitual correction. The patients experienced vision fluctuation and discomfort with their lenses. They were diagnosed with contact lens induced papillary conjunctivitis (Case1) and contact lens induced ocular surface disease (Case2) and uncorrected irregular astigmatism secondary to keratoconus (both cases). Following the management of the ocular surface disease both patients were fit into Europa scleral contact lenses which provided excellent vision and comfort to the patients.

Conclusion: Scleral lenses are indicated in the management of complex corneal and ocular surface irregularities. These lenses vault the cornea and provide crisp vision expected of a rigid contact lens as well as comfort comparable to a soft lens.

Keywords: Scleral contact lenses, Keratoconus, rigid gas permeable contact lenses, Ocular Surface disease.

Introduction

Keratoconus is a primary corneal ectatic disease; it is a non-inflammatory progressive condition of the cornea characterized by ectasia and thinning which leads to irregular astigmatism and decrease in vision[1]. Patients with keratoconus have corneal irregularities, and aberrations[2] which can be ameliorated with the use of rigid gas permeable (RGP) contact lenses[3].

The management of keratoconus varies with disease severity; although in the early stages it can be managed with spectacles or soft contact lenses, rigid gas-permeable (RGP) contact lenses (CLs) are the options of choice[4].

Scleral lenses are rigid gas permeable lenses which sit on the sclera without touching the cornea or limbus; this

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creates a reservoir between the posterior surface of the scleral lens and the anterior corneal surface. This fluid reservoir is also called clearance.

Leonardo da Vinci in 1508 conceptualized the idea of neutralizing the cornea using an enclosed fluid reservoir. This fluid reservoir between the posterior surface of the scleral lens and the anterior corneal surface corrects the irregularity on the surface of the cornea and also provides a healing surface for ocular surface disease and severe dry eyes[5].

Scleral lenses were described as early as the 1800s but their use did not achieve widespread acceptance due to complications associated with poor oxygen permeability of the materials used in its manufacture (mainly corneal hypoxia) and difficulty in manufacturing the lenses[6].

The first description of the manufacture of scleral lenses with rigid gas permeable (RGP) materials was first made in 1983 by Ezekiel[7]. This ushered in an era where scleral lenses could be used with reduced risks of corneal hypoxia and currently there is a wide range of RGP materials and designs available for scleral lenses.

The Scleral Lens Education Society (SLS) divides scleral lenses into two broad categories:

(a) Mini-scleral lenses which are scleral lenses whose diameter is up to 6mm larger than the horizontal visible iris diameter (HVID) and

(b) Large scleral lenses whose diameter is more than 6mm larger than the HVID of the cornea[5].

Scleral lenses are indicated for vision improvement in conditions characterized by high corneal irregularities like primary corneal ectasias which includes Keratoconus, Pellucid Marginal Corneal Degeneration (PMCD), Terrien's Marginal Corneal Degeneration (TMCD).

Scleral lenses are used in visual rehabilitation in cases of secondary corneal ectasias following refractive surgeries (for example, post refractive surgery ectasia and high corneal astigmatism post corneal transplantation).

Scleral lenses are also indicated in the management of severe ocular surface disease[8] as they serve the purpose of ocular surface protection and also help in promoting healing of the ocular surface and restoration of ocular surface homeostasis.

We present the visual management of 2 patients diagnosed with keratoconus using mini-scleral lenses as well as the rehabilitation of the ocular surface to achieve comfortable scleral lens wear.

Case Report

Case One

E.S, a 32-year-old Nigerian female presented to our clinic with complaints of itching in the both eyes (BE) for a month and blur vision for over 10 years in the right eye (RE). She had associated grittiness and occasional redness without discharge and had a history of eye rubbing. She is a known soft CL wearer (RE custom soft toric CL and LE spherical soft CL) for 3 years and has been wearing soft CL since the diagnosis of RE Keratoconus and left eye (LE) myopia 3 years back as well . Her current CLs were obtained 9 months back. She discontinued CL wear due to discomfort and blurred vision on the RE. She had no history of sleeping or bathing with CL on. She had used Refresh Multipurpose Solution (Optika Co. Ltd, Korea) for the daily cleaning and soaking of her CLs.

On examination, her uncorrected visual acuity (VA) was 20/200, N46 and 20/20, N5 in the right and left eye respectively, with pin-hole (PH) acuity of 20/60 in the RE.

Slit lamp examination revealed Grade 3 papillae on the upper tarsal conjunctiva (UTC) of both eyes (BE) with Fleisher's ring and Prominent corneal nerves noted in the mid periphery of the cornea of the right eye (RE). Other slit lamp findings were within normal limits. She had an intraocular pressure of 08.00mmHg and 10.00mmHg at 10:00am (Pulsair intelliPuff, Keeler Ltd, UK) in the right and left eye respectively and undilated fundus examination revealed pink optic discs with a cup to disc ratio of 0.3 with healthy neuro-retinal rims in BE. The retina was flat with the blood vessels normal in course and caliber, the foveal reflex was bright and there were no retinal abnormalities noted.

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Retinoscopy values were:
RE: -7.50/-8.75 X 170 (with RE scissors reflex) and LE:-0.25DS,
Subjective refraction was RE: -5.00/-6.00 X 180 (20/120, N46) and LE: Plano (20/20, N5).
Horizontal Visible Iris Diameter: 11.6mm in BE

Keratometry findings were:
RE: 51.50D @96 and 57.75D @6 (corneal astigmatism: -6.25DC X96) and
LE: 44.25D @70 and 44.50D @160 (corneal astigmatism: -0.25DC X70).

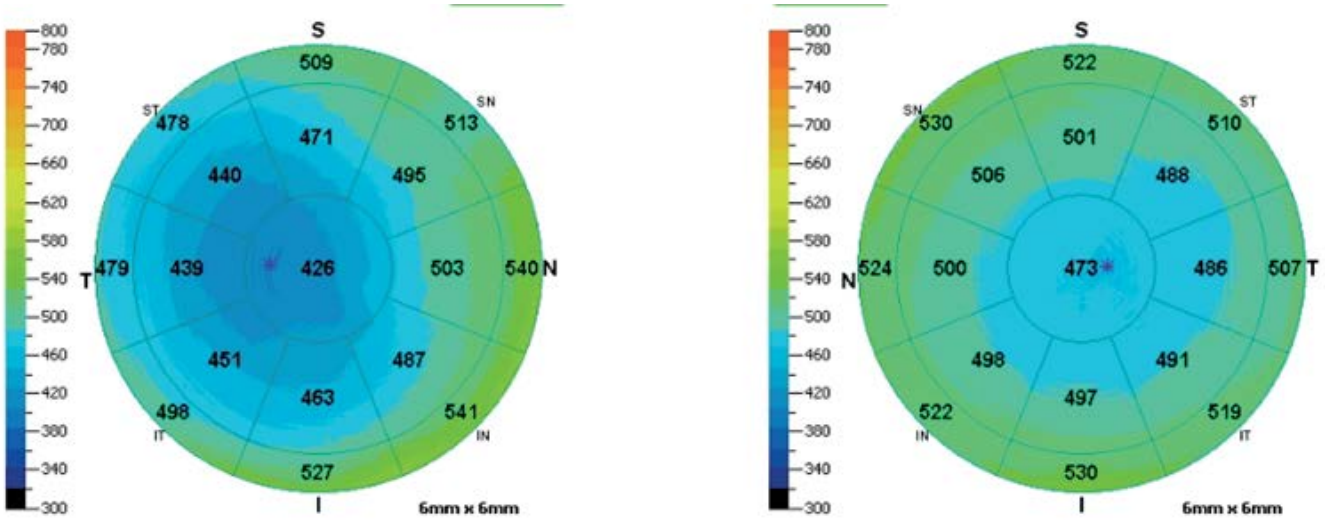

Fig1. Image of the pachymetry map in both eyes. Note: the thinnest location as indicated by the asterisk (*) is 405 microns and 463 microns in the right and left eyes respectively

The patients soft CL was inserted into BE, the CL prescription was:
RE: 8.40mm/-5.00/-4.00 X 160/ 14.50mm and
LE: 8.60mm/-0.75DS/14.00mm.
She had a VA of 20/80, N24 in the RE and 20/25, N6 in the LE with the CL on.

Due to unsatisfactory vision in the RE an attempt was made to improve the vision using a Europa scleral lens (Visionary Optics LLC, USA) with the following parameters:

Lens name and material: Europa scleral lens, Boston XO.
Design: The Europa scleral lens has one central and four progressively flattened peripheral curves divided into 3 zones (the central zone, the limbal zone and the scleral zone).
Base curve: 45.00D
Diameter: 16.00mm
Sagittal depth: 4.56 mm
Power:-1.50DS

Fit description:
After 30 minutes of adaptation following lens insertion, the lens exhibited a central clearance of 251 microns and limbal clearance of 165 microns. There was mild mid haptic compression 360 degrees with good peripheral alignment, mild impingement at 3'0 Clock over a mild nasal pingueculum.

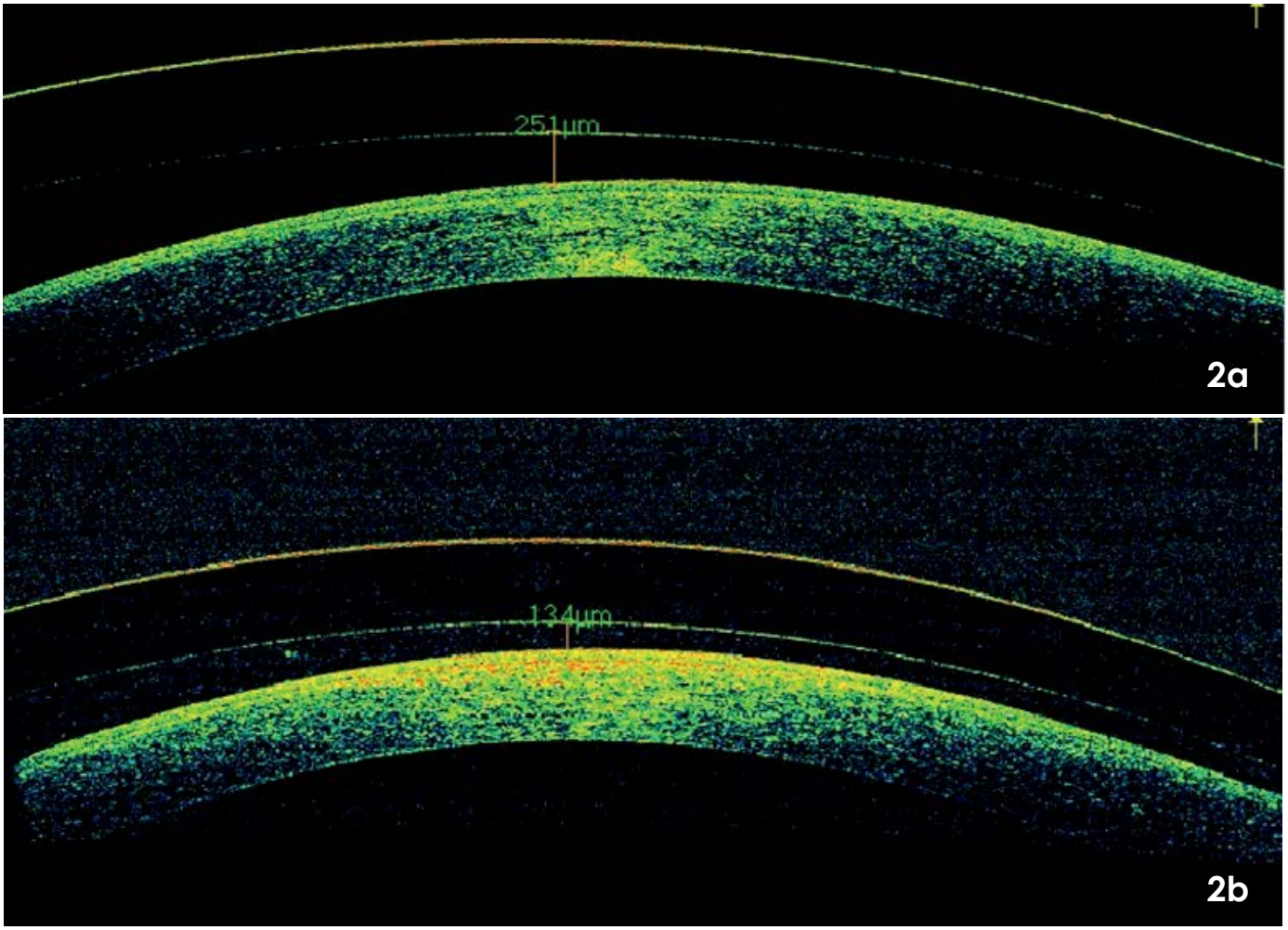

Fig2. An anterior segment –optical coherence tomography (OCT) image of the RE with lens in place, note the central clearance of 251 microns 30 minutes after lens insertion (a) and 134 microns after 4 hours of lens settling (b).

Following 4 hours of lens settling the central clearance was 134 microns and the limbal clearance was 75microns. Peripheral lens alignment was good with no blanching but mild conjunctival prolapse was still notable inferiorly.

Over refraction was +1.00DS with a visual acuity (VA) of 20/25+1, N6 in the right eye.

She was pleased with the outcome and the improved vision and comfort which she experienced with the lens in the RE.

On lens removal there was no suction, no tenderness but there was mild conjunctival prolapse inferiorly.

Lens parameters adjustment

In order to improve the lens fit and decrease the conjunctival prolapse inferiorly the diameter of the lens was increased by 0.50mm. This increase in diameter brings about an increase in the vault by approximately 100 microns. Hence in order to maintain the final tear reservoir as during trial the central base curve was flattened by 1.00D. Due to the change in base curve a compensatory change was made to the back vertex power (based on the 'SAM FAP rule').

As a result of a 1.00D flattening in the base curve, +1.00D was added to the final contact lens power making the final power: +0.50DS.

The final contact lens ordered as shown in Table 1.

Table1: Final scleral lens parameters in the RE.

Lens Parameter	Right Eye
Base curve	44.00D
Diameter	16.50mm
Sagittal height	4.86mm
Power	+0.50DS
Material	Boston XO

She was diagnosed with Right eye Contact Lens associated Papillary Conjunctivitis (CLPC), Right eye Unilateral Keratoconus with Irregular Myopic Astigmatism and Simple Myopia on the Left eye.
She was advised to discontinue CL wear in the LE as the refractive error was not visually significant and her previous LE CL had a power of -0.75DS which could cause asthenopic complaints.

Management included Sodium Cromoglycate 2% (Ivycrom, Ivee Aqua Epz Ltd. Kenya) eye drops four times daily for two weeks. Patient was advised to discontinue CL wear in BE until resolution of the conjunctivitis. She was asked to return for follow up in 2 weeks.

Dispensing visit

On her follow up visit, she reported an improved symptomatic relief and had quiet eyes with grade 1 papillae on the UTC of BE on slit lamp examination. Other findings were within normal limits. The new lens fit characteristics were similar to the last trial visit. A hydrogen peroxide based CL solution (Refine One step, Coopervision, UK) was advised for daily cleaning of the contact lenses. She was counselled about the proper lens care regimen and hygiene, and taught lens insertion and removal techniques.

Follow up visit One

On her one month follow up visit after dispensing the scleral lens, she had an average wearing time of 8 hours daily, and was adherent to all lens care regimen advised. The lens exhibited optimal central and peripheral fitting characteristics and she had a visual acuity of 20/25, N6 in the RE.
She was assessed to be doing well with the scleral lenses and was advised to continue with lens wear and gradually build up wearing time. The scleral lens care regimen was re-emphasized.

Follow up Visit Two:

On her three month follow up visit she was also doing well with the lenses and adhering to lens care regimen. She now had an average lens wear time of 12 hours daily. She was advised to follow up six monthly with the clinic.

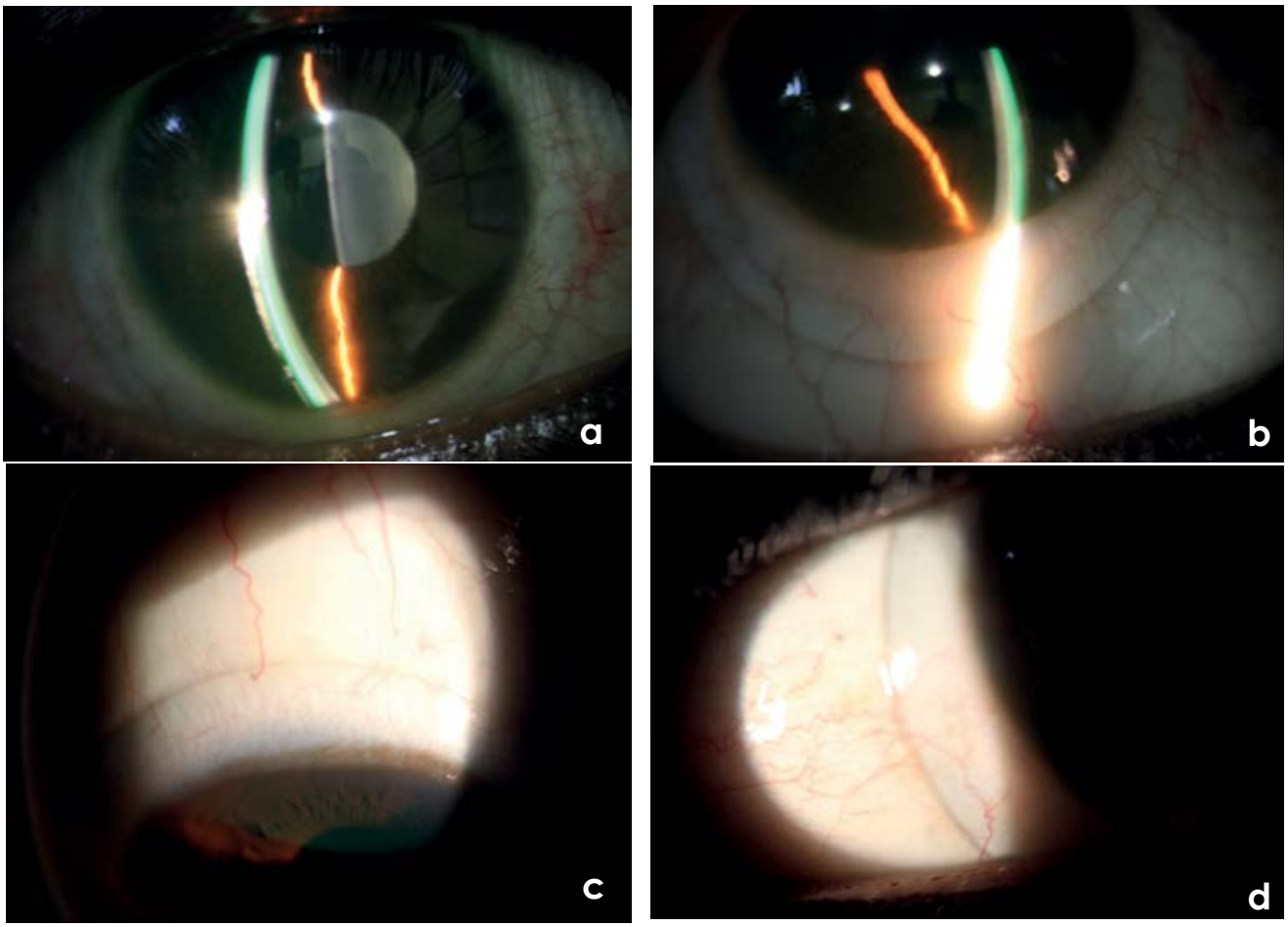

Fig3. Image of the scleral lens in the RE showing the post lens tear reservoir or central clearance ('a' and 'b') (fluorescein filled area) and the peripheral alignment fit (image 'c' and 'd').

Case Two

A.U, a 34 year old Nigerian female presented to the clinic on the 10th of March, 2018 with complaints of blur vision and discomfort with her CL in BE. She was diagnosed with bilateral keratoconus 10 years ago during a routine eye exam and had a history of CL wear for the past 10 years. She was currently wearing a custom soft toric CL in the RE which was 4 months old and a Hybrid lens in the LE which was 1.5 years old. She had an average CL wear time of 16 hours daily and complained of discomfort worse at the end of the day. She was currently using a hydrogen peroxide solution for daily cleaning of the hybrid CL and a multipurpose solution for the daily cleaning and soaking of the custom toric soft CL.

Aided VA (with CL on) was $20/50^{+1}$; N10 and $20/30^{-2}$; N8 in the right and left eyes respectively. Slit lamp examination revealed few blocked meibomian orifices on the upper and lower lid margins, moderate conjunctival congestion and the CL surface in BE was remarkable for deposits and scratches on the surface of the rigid portion of the hybrid lens. The hybrid CL in the LE had cracks at the junction between the rigid and the soft lens skirt.
On lens removal sterile sub-epithelial corneal infiltrates and mild superficial staining in the nasal and temporal mid periphery in the RE was noted as well as and an indentation ring in the LE (due to impingement from the rigid portion of the hybrid CL) with 360 degree limbal redness.

On ocular surface evaluation her Ocular Surface Disease Index (OSDI) questionnaire score was 29.16 (Moderate dry eye symptom) and she had Tear Break Up Time (TBUT) of 3 seconds in BE, Tear meniscus height of <0.2mm and Schirmer's (with anesthetic) of 15mm and 20mm in the right and left eyes respectively after 5 minutes of measurement. On expression of the meibomian glands with mild pressure on the lids there was cloudy meibum secretion from most of the orifices of the upper and lower lids.

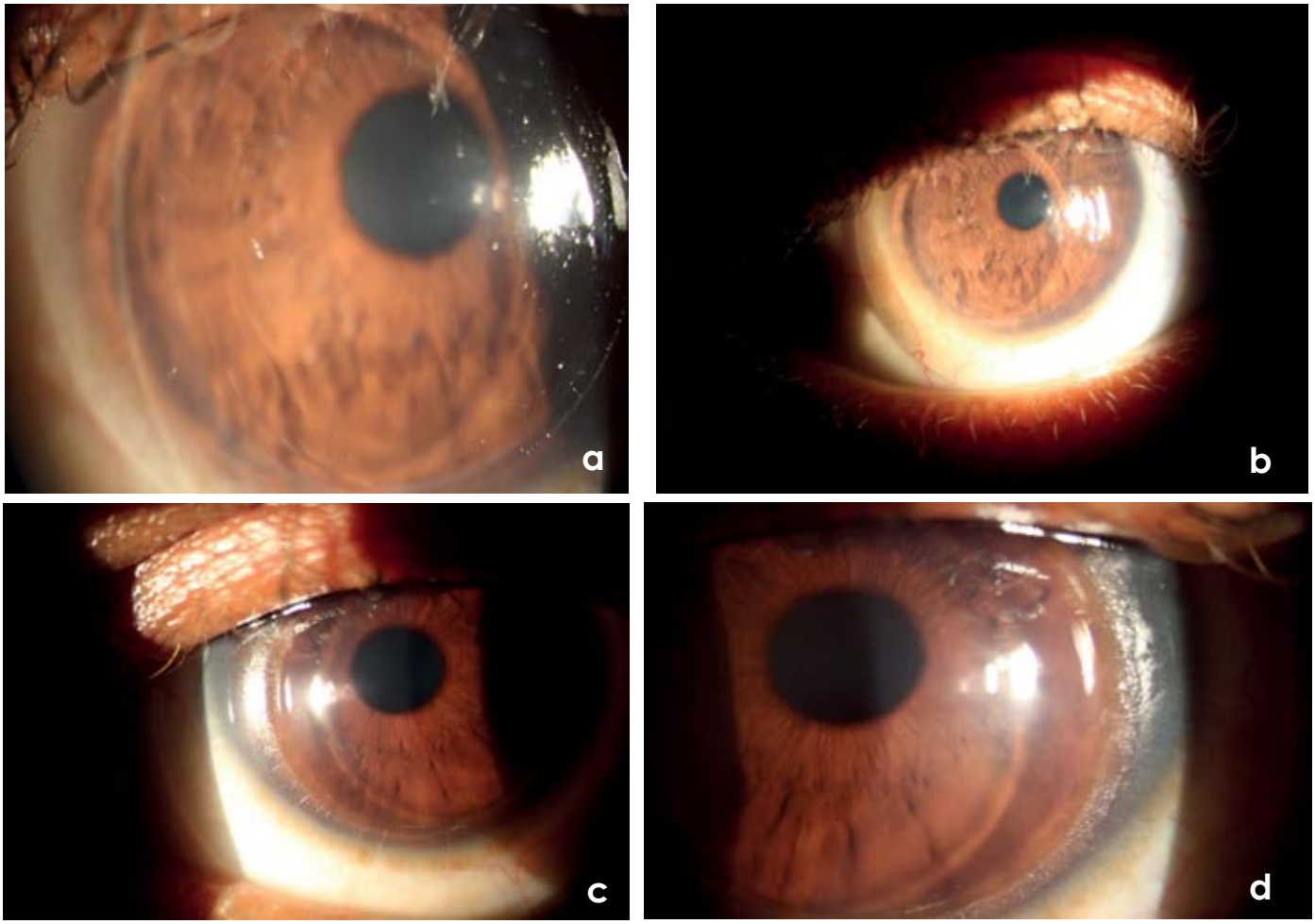

Fig4. Image of the soft toric CL in the RE and hybrid CL in the LE. Note the surface deposits (a) and temporal decentration of the lens (b); also note the deposits on the hybrid lens at the junction of the soft and rigid lens (d) and cracks on the soft lens skirt (c).

She was diagnosed with Contact Lens Induced Meibomian Gland Dysfunction with Evaporative Dry Eye and Asymptomatic Infiltrative Keratitis in the RE. She was advised to reduce her CL wear time, and discontinue for at least a week prior to her next visit in order to allow for normalization of corneal parameters before the next visit. She was counselled to use hydrogen peroxide solution (Refine One Step, Coopervision, UK) for cleaning of both lenses to decrease the likelihood of preservative induced ocular toxicity and dry eyes and she was counselled to perform warm compress, lid massage and lid scrubs twice daily. She was advised Fluorometholone 1% eye drops four times daily for a week and twice daily for a week in the RE and she was also prescribed artificial tear drops (Hypermellose 0.3%) four times daily for a month in BE.

Follow up Visit one

She returned in 2 weeks feeling better, she presented without her contact lenses on as advised. She had an unaided VA of 20/400, N48 and 20/400, N24 in the right and left eyes respectively. On slit lamp examination there were no sub-epithelial infiltrates and the eyes appeared quiet and intraocular pressures were 12.0mmHg at 12:00pm in BE (Pulsair tonometer, intelliPuff, Keeler Ltd.)

Retinoscopy revealed:
RE: -15.00DS/-5.25DC X 15
LE: -12.00DS/-9.50DC X 160 (Scissors reflex was noted in both eyes)

Subjective acceptance:
R: -12.00DS/-4.00 X 170 (20/70; N12)
L: -11.00DS/-4.00DC X 170 (20/70; N12)

Keratometry readings were:
R: 50.75 @ 160 and 54.44 @ 70
L: 47.07 @ 16 and 52.49 @106

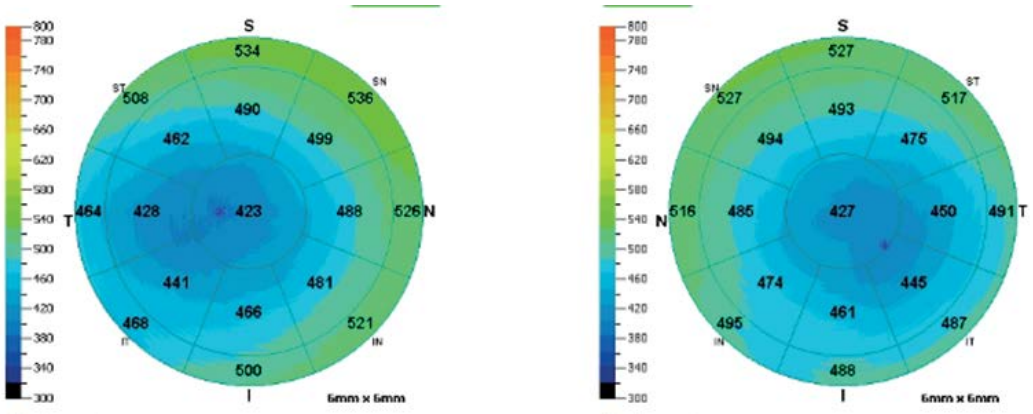

Fig5. Image of the cornea pachymetry map in both eyes. Note: the thinnest location as indicated by the asterisk (*) is 404 and 410 microns in the right and left eyes respectively

The scleral lens trial was advised. Europa scleral lenses were tried with the following parameters:

Table2. Parameters of the inserted scleral lenses

Lens Parameter	Right Eye	Left Eye
Base curve	7.50mm	7.67mm
Diameter	16.00mm	16.00mm
Sagittal height	4.56mm	4.47mm
Power	-1.50DS	-1.00DS
Material	Boston XO	Boston XO

After 30 minutes the fit was assessed and there was a central clearance of approximately 500 microns in each eye with a limbal clearance of approximately 200 microns. There was good peripheral alignment and no impingement; no blanching of vessels, there was mild mid haptic compression in both eyes.
She was instructed to continue to wear the lenses for at least 4 hours.

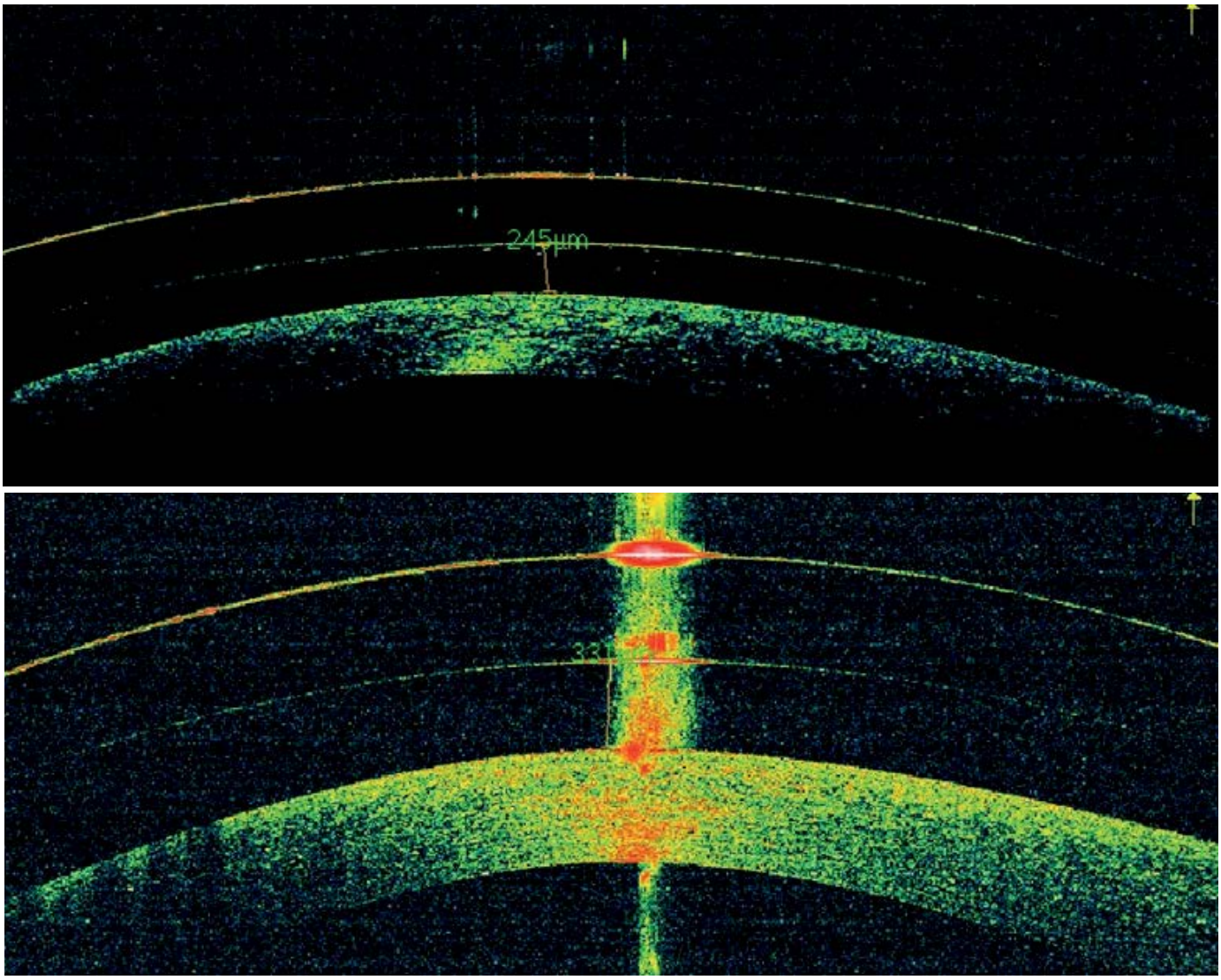

Fig6. The central corneal clearance was 245 microns in the RE and 331 microns in the LE with a limbal clearance of 100 and 190 microns in the right and left eyes respectively after 4 hours of lens settling.

Over refraction with the lenses was:
RE: -5.00DS (20/30-1, N8) and LE: -5.50DS (20/20-2, N8) and BE: 20/20, N6.
On lens removal there was no suction, no tenderness but there was mild limbal microcystic edema more in the limbal area in the LE. She was comfortable with the scleral lenses in both eyes and was willing to proceed with the lens order.

In an attempt to improve the VA in the RE and decrease the perceived corneal aberrations in the eye, the central base curve of the RE lens was flattened while leaving other parameters same so as to bring the lens closer to the central cornea. In the LE in order to decrease the limbal edema, the peripheral curve (PC) 2 and 3 were flattened by 0.50mm which decreased the overall clearance by 100 microns but to maintain the central base curve and central clearance, the central base curve was steepened by 1.00D with the necessary power adjustments made.

The final contact lens prescribed was:

Table3. Final scleral lens parameters for Case 2

Lens Parameters	Right Eye	Left Eye
Base curve	7.67mm	7.50mm(PC2:9.50mm and PC3: 13.50mm)
Sagittal height	4.524mm	4.453mm
Power	-5.25DS	-7.25DS
Diameter	16.0mm	16.0mm
Material	Boston XO	Boston XO

Lens dispensing visit

The patient presented for lens collection 2 weeks later. The lens exhibited similar fitting characteristics similar to the last trial visit. There was a central clearance of approximately 300 microns in BE and a limbal clearance of about 100 microns, the lens exhibited a good peripheral alignment with no blanching or impingement of conjunctival blood vessels.

She was taught lens insertion and removal techniques and advised to continue with the hydrogen peroxide based solution for daily cleaning of her contact lenses. She was advised to adhere strictly to lens care regimen to avoid complications and to use the preservative free lubricating drop over the contact lens as needed.

Follow up visit two:

The patient presented a month after her lens dispensing visit. She was comfortable with the lenses and reported an average wearing time of 12-13hrs. She was compliant with CL wear regimen and care.

She had a VA of 20/20-2 in each eye and N5 at near with the scleral lenses; and over refraction was Plano in BE. On lens removal there was no corneal or conjunctival staining, no tenderness and no suction on lens removal. She was advised to continue with her Europa scleral lenses and continue with same lens care regimen. The patient was doing successfully well with scleral lenses up to her last follow up 6 months after the initial dispensing visit.

Discussion

The prevalence of keratoconus differs based on the geographic location, criteria used in definition of keratoconus, and the study population[9]. There are no studies known to the researchers about the prevalence of the disease in Nigeria, and there is limited data regarding its prevalence in black African population. A study highlighting the characteristics of keratoconus patients in a Kenyan contact lens practice reviewed the ophthalmic records of 254 patients and reported that 75% of patients with keratoconus were between the ages of 6-25years and 71% had severe keratoconus (this study classified keratoconus based on the Collaborative Longitudinal Evaluation of Keratoconus study criteria)[10].

The age of onset of keratoconus occurs in early teenage years to early adulthood. The disease goes on to progress for about 20 years after onset[11]. Though age is theorized to have a protective effect on keratoconus owing to natural stiffening of the collagen fibers with age[12], in order to decrease the progressive thinning and ectasia of the cornea and avoid increased irregular astigmatism and decreased vision associated with the disease, corneal collagen cross linking procedure has emerged as the surgical option of choice in decreasing or halting progression of the disease[13]. Our patients presented in their third decades but the self-reported age of onset was still after the teenage years in both patients. It is also possible that the onset of the disease occurred earlier but the patients delayed in seeking ophthalmic consultation. Keratoconus is known to affect both genders though some studies report a male preponderance[14] and others report higher female prevalence[15].

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There are varying etiologies of keratoconus, the most common associated risk factor for developing keratoconus is eye rubbing, and other possible etiologies of keratoconus include atopy, genetics[16] and family history[17]. The patient in case 1 had a history of eye rubbing.

As the disease progresses there is a need for rigid gas permeable contact lenses[4]. Scleral lenses provide excellent vision expected of a rigid gas permeable contact lens as well as comfort similar to a soft contact lens because they rest on the conjunctiva which is not as sensitive as the cornea.

Scleral lenses have been used even in severe stages of keratoconus where corneal transplantation is indicated and have been shown to decrease the need for corneal transplantation[18]. In fitting of scleral lenses the tear reservoir created behind the lens (clearance) provides an environment to correct the anterior corneal irregularity and the over-refraction incorporated into the lens power corrects the residual refractive error. As noted in our patients with high corneal astigmatism secondary to keratoconus, visual acuity improved remarkably in these patients after scleral lens wear.

As with other contact lens modalities, complications have been known to occur with scleral lens wear which include but is not limited to hypoxic events mostly due to an excessive clearance, discomfort due to poor fitting relationship between the lens periphery and the sclera on which it sits, mid-day fogging due to the collection of debris in the tear reservoir or a flat fitting scleral lens and infections due to poor compliance with lens care regimen[19].

Conclusion

Scleral lenses are useful in the visual rehabilitation of keratoconus and other corneal ectasias even in very advanced stages. The benefit of this contact lens modality goes beyond just visual recovery as it also aids in ocular surface rehabilitation. The use and awareness of scleral lenses in Nigeria is still in its infancy but eye care practitioners should avail this option to patients when indicated.

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