



Scleral Lens Fitting in a Patient with Congenital Peter's Anomaly

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Abstract

Peter's anomaly is a congenital ocular malformation that optometrists rarely encounter. Proper assessment of cornea and anterior segment has an important role in best lens selection with the best visual outcome possible. This case report outlines a discussion of scleral lens fitting in Peter's anomaly.

Keywords: Peter's Anomaly; Scleral Lens

Introduction

Peters' anomaly is a very rare autosomal recessive ocular malformation that can be associated with systemic abnormalities. Systemic abnormalities include: cleft lip and palate, facial dysmorphism, syndactyly, brachycephaly, as well as cardiac, neural, and hearing deficits [1]. The incidence and prevalence of Peters' anomaly internationally remains unknown. Peters' anomaly can be classified into two subtypes: Type 1, and Type 2. In Type 1, 80% of cases present bilaterally with central and paracentral corneal opacification. The cornea is typically avascular; iris strands extend from the collarette, and systemic abnormalities are not usually present. In Type 2, cases are commonly bilateral with denser corneal opacification; there is often juxtaposition of the lens, and iris strands may or may not be present. The posterior stroma and Descemet membrane is classically malformed. The incidence of systemic abnormalities is more common in Type 2 [1].

Sensory deprivation, amblyopia, and glaucoma are significant sequelae of Peters' anomaly [2]. The eyes with Peters' anomaly can be classified into three groups according to the treatment protocol: (1) the medical treatment group, (2) the surgical treatment group, and (3) the no-treatment group. Medical treatment comprises of occlusion therapy with or without pupil dilation with mydriatics. If the media opacity is too dense to observe the fundus, various addi-

tional surgical treatments are performed. Cases in which no treatment was available were placed in the no-treatment group.

Mild disease has visual acuity of 20/100 or better, while eyes with severe disease has visual acuity categorised as finger-counting (CF) or worse [1].

Corneal scarring leads to a decline of visual acuity either by light scatter due to an irregular cornea or by direct obscuration of rays [4]. Scleral lenses can be a great option for many patients with corneal scars and corneal degenerations or dystrophies since it improves visual acuity by creating a smooth anterior optical surface [3].

Case Report

Initial Visit, Brief Eye Exam, 06/06/2022

A twelve-year-old female, born full term to a non-consanguineous marriage presented for a eye exam accompanied by her mother with the complaint of poor vision since childhood. Her chief complaints were not being able to recognize people's face and difficulty reading and writing.

The patient's last eye exam was a comprehensive eye exam some six years back at another clinic. This was the first time she pre-

sented to our clinic. There was a positive family history of peters anomaly in her maternal lineage. The patient denied any history of ocular surgeries or trauma. She was not taking any medications and had no known drug allergies. She was oriented to time, place, and person, and her mood was appropriate, albeit noticeably anxious and nervous about the examination.

The patient's uncorrected distance visual acuity was 1/60 and 2/60 in the right and left eye respectively. She had no habitual spectacle correction. Function of extraocular muscles were found to be smooth, accurate, full, and extensive with no pain. Unilateral and alternating cover testing revealed hypertropia of RE at distance and near.

Examination of the anterior segment revealed several remarkable findings. Eyelids were clean and clear, without lesions. The bulbar conjunctiva in BE were transparent and equally lustrous. There were bilateral central and paracentral corneal opacities at the level of posterior stroma in both eyes (Right eye being more severe than the left eye). In RE, underlying iridocorneal synechiae were present that extended from the iris collarette to the border of the corneal opacity, which was further confirmed by the AS OCT (Figure 3). The iridocorneal synechiae were absent in the LE. The sclera was clear in each eye (Figure 1,2).

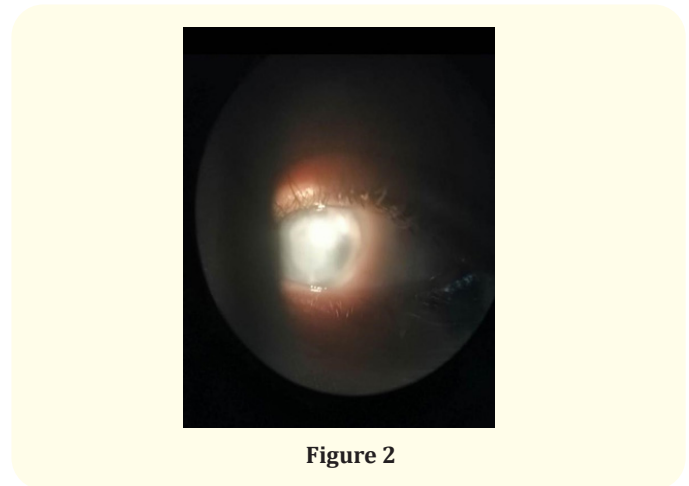


Figure 2

Differential diagnoses considered at this point include: Axenfeld-Rieger anomaly and syndrome, iridocorneal endothelial syndrome, posterior polymorphous dystrophy, congenital hereditary endothelial dystrophy, and congenital hereditary stromal dystrophy.

However, in lieu of the physical examination revealing dense bilateral cataracts, bilateral iridocorneal adhesions in conjunction with central and paracentral corneal opacification at stroma with cornea being avascular and iris strands extending from the collarette with no other systemic abnormalities, these findings are consistent with Peters' anomaly (type I).

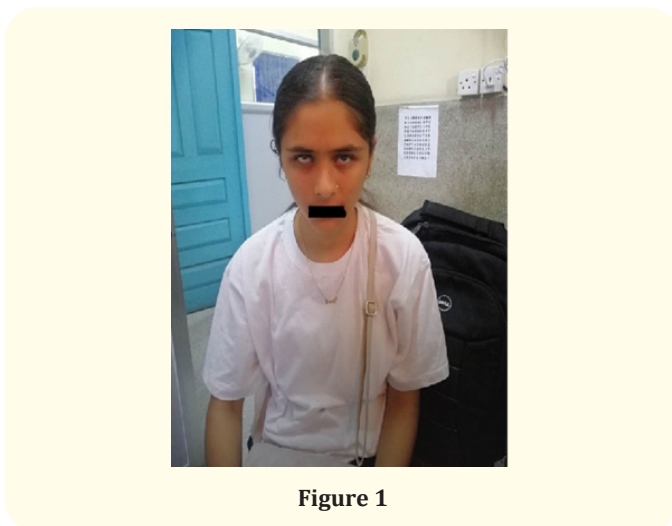


Figure 1

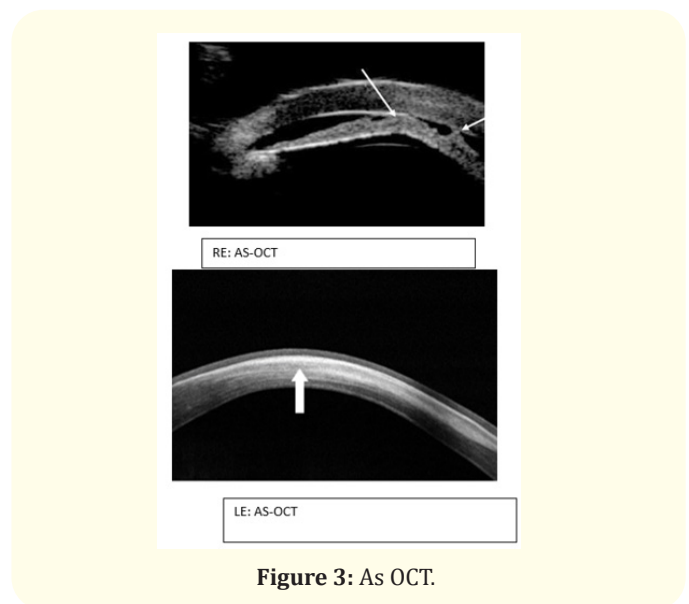


Figure 3: As OCT.

The patient was then asked to make a follow up visit in the contact lens clinic for the potential visual rehabilitation with scleral contact lens. Follow-up #2, Scleral lens trial, 12/06/2022.

Before trial

Eye	Unaided Visual acuity	PIN HOLE Visual acuity	K-READING
OD	1/60	1/60	7.65mm/7.74mm
OS	2/60	6/36	7.6mm/7.7mm

Table 1

Scleral lens trial

Eye	Parameters	Over the lens acceptance	Visual acuity
OD	7.70/L3/3.98 -2.00/14.50		1/60(NI)
OS	7.50/L3/4.13 -2.00/14.50 (vaulting↑)	+11.00	3/60

Table 2

Owing to the dense stromal opacities, the visual acuity of the right eye was not improving even after multiple trials. So, we only included the recording of LE, the trial of which were eventful.

Scleral trial 2

Eye	Parameters	Over the lens acceptance	VA
OS	7.70/L3/3.98 -2.00/14.50 (vaulting was acceptable)	+10.00	6/60 (fluctuating)

Table 3

Scleral trial 3

In order to reduce the aberration of Spectacles (+10.00D) in over-refraction, mini scleral lens with parameters nearly equal to that of scleral trial #2 along with +10.00 BVP was tried.

Eye	Parameters	Over the lens acceptance	VA	Vaulting (after 30 min)	Blanching (30 min)
OS	7.70/L3T/4.54/+10.00/16.00	+0.50	6/36	Acceptable	Absent

Table 4

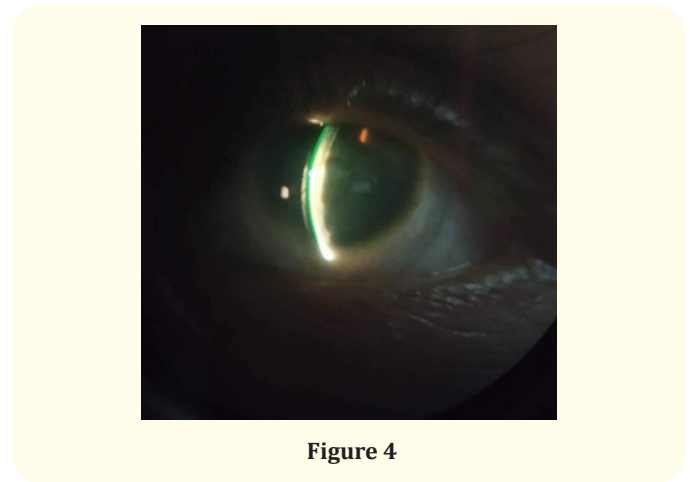


Figure 4

Discussion

Scleral lenses applied with preservative-free fluid in the post-lens-fluid reservoir, continuously bathes the ocular surface and corrects for corneal irregularities—improving visual acuity by creating a smooth anterior optical surface [3].

Scleral lenses are particularly helpful for conditions like corneal transplantation degenerations or dystrophies such as Salzmann’s nodular degeneration or Terrien’s marginal degeneration because they correct irregular astigmatism after corneal transplantation, lessen corneal scarring over time, correct corneal irregularities by vaulting over peripheral nodules in Salzmann’s and decrease the amount of corneal neovascularization while continuously bathing the ocular surface during wear [3].

For corneal scars causing opacity and corneal irregularity along the visual axis, scleral lenses can create a smooth surface and improve vision [3].

In a developing country like ours where corneal scarring accounts for more than 1/5th of the total burden of blindness and good corneas are in short supply, any suitable alternative to keratoplasty is welcomed [5]. Research has shown improvement in BCVA by three or more decimal acuity lines in cases like, Keratoconus, Interstitial keratitis, Herpetic Keratitis, Stevens Johnson syndrome and acid Burn [4]. Scleral lens can be a solution in patients waiting for keratoplasty. A study by Smiddy, *et al.* found that 69% of patient who were referred for keratoplasty could be successfully fitted with contact lenses delaying the need for surgery [6].

Conclusion

Scleral lenses can be a great option for many patients with corneal scars and corneal degenerations or dystrophies. With the right pre-fit testing and with a firm understanding of the potential fitting challenges we can successfully fit scarred and irregular cornea with scleral lenses. They can go a long way to improve patients' quality of life, visual acuity and ocular comfort.

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