

## Recurrence or Re-emergence of Keratoconus – What is the Evidence Telling Us? Literature Review and Two Case Reports

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**ABSTRACT** Keratoconus may recur following penetrating or lamellar keratoplasty, but latency is considerably longer in the former. Since keratoplasty involves only partial excision of the cornea, and recent research strongly indicates the presence of the pathology in the peripheral host cornea, the reappearance of the pathology after a latency period is most likely due to migration of the disease from host to donor cornea. This notion is further corroborated by the shorter latency period in partial thickness keratoplasty, where more of the diseased host cornea remains in place. Other proposed causes for the recurrence of keratoconus, such as eye rubbing and contact lens wear, were reportedly not associated with a significant number of cases, and, therefore, are not the primary factor. Based on existing literature, it is concluded that, in post-keratoplasty keratoconus, the etiology stems from re-emergence of the disease rather than recurrence. Keratoconus patients in need of keratoplasty should be counseled on the possibility of the disease re-emerging.

**KEY WORDS** ectasia, etiology, eye-rubbing, histopathology, keratoconus, post-keratoplasty keratoconus, recurrence latency, recurrent keratoconus

Accepted for publication May 2014.

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Sources of support: None.

The authors have no proprietary or commercial interests in any concept or product discussed in this article.

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© 2014 Elsevier Inc. All rights reserved. *The Ocular Surface* ISSN: 1542-0124. Bergmanson JPG, Goosey JD, Patel CK, Mathew JH. Recurrence or Re-emergence of Keratoconus – What is the Evidence Telling Us? Literature Review and Two Case Reports. 2014;12(4):267-272.

### I. INTRODUCTION

**I**nfrequently, literature reports remind us that keratoconus can recur in the recipient of a corneal transplant. Rabinowitz suggested that the recurrence may be due either to the existence of the disease in the remaining host tissue or to the presence of subclinical disease in the donor cornea.<sup>1,2</sup> Maguire believed that most recurrences resulted from incomplete cone excision.<sup>3</sup> The rare recurrence of keratoconus was also recognized by Leibowitz and Morello, who proposed that postoperative contact lens wear may contribute to the return of corneal ectasia but is not the only factor,<sup>4</sup> as this complication was also reported in a patient who had not worn contact lenses.<sup>5</sup>

To enhance evidence-based understanding of the etiology and causes of recurrent keratoconus, we present two case reports of bilateral recurrent keratoconus and review reports from the peer-reviewed literature on this complication of corneal transplantation. Our findings suggest that *re-emergent* keratoconus may be a more appropriate term than *recurrent* keratoconus to describe the reappearance of this pathology after corneal transplantation.

### II. CASE REPORTS

#### A. Case Report #1

A 52-year old white male was referred to the University of Houston, University Eye Institute Cornea and Contact Lens Service for a second opinion on suspected recurrent keratoconus and a contact lens evaluation. The patient had previously undergone bilateral penetrating keratoplasty (OD, Mar 1990; OS, Aug 1990) due to progressive keratoconus. The patient denied a history of excessive eye rubbing and reported alternating between hybrid (gas permeable/soft) contact lenses and spectacles for the past few years. On initial examination, he had a visual acuity of 20/25 OD and 20/400 OS with his usual spectacle correction but was correctable with contact lenses to 20/20 OD and OS. Biomicroscopic examination revealed clear, well-centered corneal grafts measuring 9 mm with no invading neovascularization OU and excellent endothelial mosaic on specular

**OUTLINE**

- I. Introduction
- II. Case Reports
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  - B. Case Report #2
- III. Method of Literature Review
- IV. Discussion
- V. Summary and Conclusions

endothelial reflection. Vogt’s striae, Fleischer’s ring, and Munson sign were absent. At this visit, keratometric readings were 45.62/56.49X020 OD and 40.37/59.56X170 OS. Topographic imaging with Pentacam HD (Oculus, Inc., Arlington, WA, USA) revealed bilateral irregular astigmatism with marked inferior corneal steepening (Figure 1). Pachymetric measurements with Pentacam HD revealed a thinnest locale of 458 μm OD and 494 μm OS. Based on these findings, the patient was diagnosed with apparent recurrent keratoconus OU. Repeat keratoplasty was not indicated at this time because the patient was correctable to 20/20 OD and OS. The patient was advised to return to our clinic for scleral contact lens fitting OU and to visit the referring practitioner for continued monitoring of corneal status. Histopathological confirmation of keratoconus was not feasible in this case.

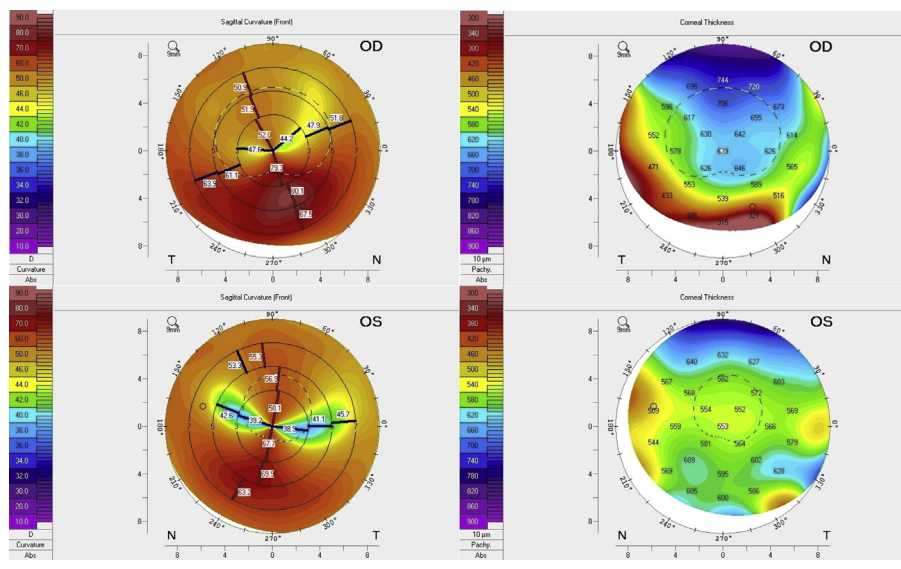
**B. Case Report #2**

A 58-year-old Hispanic female was referred to the University of Houston, University Eye Institute Cornea and Contact Lens Service for re-evaluation of her habitual contact lenses. The patient had been fitted with scleral gas permeable contact lenses OU approximately 2.5 years earlier. Prior to that, the patient wore corneal gas permeable contact lenses with limited success. Review of this patient’s medical history revealed a prior diagnosis of keratoconus

and numerous ocular surgeries: penetrating keratoplasty (OU 1981, 6 months apart), cataract surgery with intraocular lens implantation and limbal relaxing incisions (OD, 4/28/2004; OS, 4/24/2004), photorefractive keratectomy (OU, 8/17/2004), repeat photorefractive keratectomy (OU, 4/22/2005), and YAG laser capsulotomy (OD, 4/21/2005; OS, 2/16/2007). She denied a history of vigorous eye rubbing prior to or following her initial keratoplasty in 1981. With her habitual contact lenses, visual acuities were 20/25 OD and OS. Corneal biomicroscopic evaluation revealed clear, well-centered corneal grafts measuring 8mm with no invading neovascularization OU and normal endothelial mosaic. Vogt’s striae, Fleischer’s ring and Munson sign were absent. Keratometric readings were 51.90/58.60x045 OD and 58.90/65.20x123 OS. Topographic imaging with Pentacam HD (Oculus, Inc.) revealed severe irregular astigmatism with marked inferior corneal steepening OU (Figure 2). Pachymetric measurements with Pentacam HD (Oculus, Inc.) revealed a thinnest locale of 296 μm and 151 μm in the right and left eye, respectively. Although we were unable to confirm the diagnosis histopathologically, this patient was diagnosed with apparent recurrence of keratoconus OU, which may have been caused by multiple refractive surgical procedures. Re graft surgery was not indicated, as the patient achieved visual acuity of 20/25 in each eye and had good tolerance of scleral contact lenses.

**III. METHOD OF LITERATURE REVIEW**

We considered all, to our knowledge, peer-reviewed literature on recurrence of keratoconus (Table 1) using the PubMed search engine. Search terms used were: *recurrent keratoconus, recurrent ectasia, complications in keratoconus grafts, corneal transplant ectasia, corneal graft keratoconus*. Our two new cases of post-graft recurrent keratoconus are also included in Table 1.



**Figure 1.** Topography and pachymetry (Pentacam HD) of Case 1.

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