·Letter to the Editor·

Phacoemulsification in a rare case of keratoconus with Fuch´s endothelial corneal dystrophy

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Dear Sir,

I am Dr. Jaya Kaushik from the Department of Ophthalmology of the Post Graduate Institute of Medical Education and Research, Chandigarh, India. I write to present a case report of phacoemulsification in a rare case of cataract associated with keratoconus and Fuch's endothelial corneal dystrophy (FECD).

Keratoconus with FECD is well described in literature [1,2]. Missense substitution in ZEB 1 protein is linked with keratoconus and FECD in a recent study by Lechner *et al* [3]. This study further revealed the dysregulation of α-type IV collagen which is a common link between ZEB 1 mutation and clinical phenotype (keratoconus, FECD and posterior polymorphous corneal dystrophy). The both coexisting corneal diseases pose a great challenge for cataract surgery as corneal pachymetry is less reliable to assess the severity of disease. Currently, there are no published guidelines for cataract surgery in presence of coexisting both keratoconus and FECD. In this case report we describe a good outcome in a female with keratoconus and FECD who underwent phacoemulsification and posterior chamber intraocular lens (PCIOL) implantation.

A 60-year-old female presented with complaints of blurring of vision for 1y. Ocular examination revealed best corrected visual acuity (BCVA) 20/200 in both eyes (OU), and features of keratoconus with endothelial guttae, deep anterior chamber, cataract with normal intraocular pressure (OU) (Figure 1A, 1B). The anterior chamber depth was 3.21 mm (Pentacam) in right eye (OD). Fundus details were not visible because of the dense cataract in both eyes. Pentacam (Oculus) of both eyes revealed inferior steepening consistent with keratoconus. Based on clinical parameters and investigations, patient was diagnosed to have bilateral keratoconus grade II (Pentacam^{R} derived Amsler-Krumeich stage)[4] with FECD grade IV [5,6] and corticonuclear cataract grade V (Lens Opacity Classification System III)[7].

Patient was planned for phacoemulsification with PCIOL implantation in the right eye. Her preoperative work-up included biometry (IOL Master V.5 Carl Zeiss Meditek) which revealed OD axial length 29.40 mm, keratometry K1: 50.45 D (Diopters), K2: 55.88 D, cylinder -5.43 D×26, IOL power -6 D; and pachymetry of 337 μm. Her endothelium cells (Specular Microscope SP-3000P) were 1631 in OD.

Patient underwent phacoemulsification (Alcon Infinity^{R} vision system) with -5.0 D Acrysof multi piece IOL implantation in OD. Surgical steps for phacoemulsification included 2.2 mm clear corneal incision at steeper axis, use of cohesive underneath dispersive to coat endothelium, use of torsional power around 50%, and limbal relaxing incision at the end of the surgery to minimize astigmatism. No intraoperative complications were reported. In the setting of cataract surgery alone, the possibility of postoperative corneal oedema, corneal decompensation and need for penetrating keratoplasty in future was discussed thoroughly with the patient.

On the first postoperative day, patient had mild corneal oedema. Neither ocular inflammation nor infectious complications were reported in the operated eye of the patient during follow-up. Her corneal oedema gradually resolved from postoperative first to six weeks with BCVA improved to 20/50 (Figure 1C, 1D). At last follow-up (3mo) BCVA reported was 20/40 with refractive error of -1.50/-0.50×90 and pachymetry of 393 μm OD.

Cataract surgery in patients with keratoconus and FECD poses a significant challenge as both diseases can mask each other. Jurkunas and Azar [8] published a retrospective case series and reported the potential complications of cataract surgery in patients with coexistent keratoconus and FECD: 4 out of 5 eyes developed corneal oedema/ectasia post cataract surgery. Preoperative CCT was available in only one case with 666 and 649 μm in OD and OS respectively, and preoperative endothelium cell counts were not performed. However, this case series showed the corneal pachymetry as

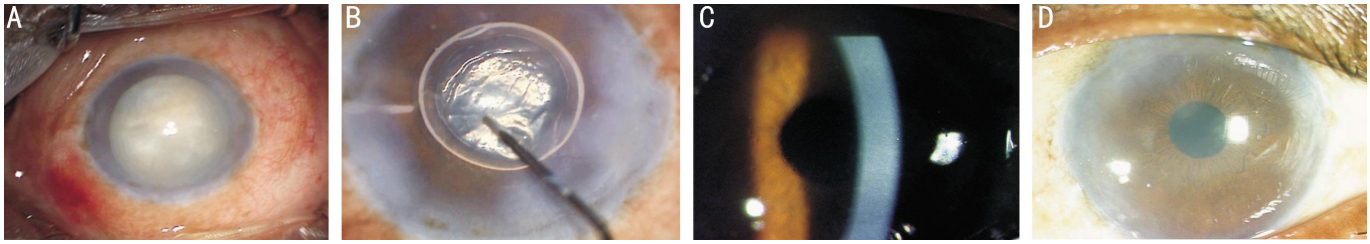

Figure 1 Clinical photographs of patient undergoing phacoemulsification in keratoconus with Fuch´s endothelial corneal dystrophy A: Intraoperative photograph of right eye showing cataract; B: Central guttae over the posterior surface of cornea; C: Postoperative (1wk) slit-lamp photograph demonstrates the vertically oriented stromal lines at the apex of the cone as seen in keratoconus and guttae can be seen just below and to the right of the lines; D: Postoperative (6wk) diffuse illumination photograph showing minimal corneal haze, well centred pupil and posterior chamber intraocular lens (IOL).

less reliable measure to assess the severity of both the diseases and recommends the routine use of preoperative corneal topography and specular microscopy in such cases. In our case, it was planned to go ahead with cataract surgery alone as the visual rehabilitation with keratoplasty is associated with multiple issues like prolong visual recovery, multiple hospital visits, management of postoperative astigmatism, suture removal and potential graft rejection. Use of low torsional power along with meticulous use of dispersive as well as cohesive agent was the key factor to reduce the endothelium cell count loss during the phacoemulsification in our case. One diopter of postoperative myopia was targeted to compensate for hyperopic shift that can occur in the event of future Descemet stripping endothelial keratoplasty [9]. Hydrophobic acrylic multipiece IOL was implanted in view of more uveal and capsular biocompatibility [10]. Recommendation for cataract surgery alone in FECD without ectasia has expanded in patients with preoperative pachymetry of 600 μm to 640 μm with current surgical methods, earlier for which (>600 μm) triple procedure (keratoplasty with cataract extraction and IOL implantation) was recommended [11]. Till now, no such guidelines reported for the phacoemusification in patients with the co-existent keratoconus and FECD.

This case report highlights the importance of comprehensive ophthalmic evaluation especially corneal topography and specular microscopy, to plan for further line of management in such cases. With current modified surgical protocol, good visual outcome can be expected in phacoemulsification and PCIOL implantation for mature cataract with coexisting keratoconus and FECD, where postoperative corneal oedema/ectasia and need for corneal transplantation is the usual expected outcome. It highlights a need of prospective interventional study showing the outcome and guidelines for phacoemulsification in patients with coexistent keratoconus and FECD in near future.

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REFERENCES

1 Lipman RM, Rubenstein JB, Torczynski E. Keratoconus and Fuchs' corneal endothelial dystrophy in a patient and her family. *Arch Ophthalmol* 1990;108(7):993-994

2 Orlin SE, Raber IM, Eagle RC Jr, Scheie HG. Keratoconus associated with corneal endothelial dystrophy. *Cornea* 1990;9(4):299-304

3 Lechner J, Dash DP, Muszynska D, Hosseini M, Segev F, Georges S, Frazer DG, Moore JE, Kaye SB, Young T, Simpson DA, Churchill AJ, Heon E, Willoughby CE. Mutational spectrum of the ZEB1 gene in corneal dystrophies supports a genotype-phenotype correlation. *Invest Ophthalmol Vis Sci* 2013;54(5):3215-3223

4 Kanellopoulos AJ, Asimellis G. Revisiting keratoconus diagnosis and progression classification based on evaluation of corneal asymmetryindices, derived from scheimpflug imaging in keratoconic and suspect cases. *Clin Ophthalmol* 2013;7:1539-1548

5 Krachmer JH, Purcell JJ Jr, Young CW, Bucher KD. Corneal endothelial dystrophy. A study of 64 families. *Arch Ophthalmol* 1978;96 (11): 2036-2039

6 Gottsch JD, Sundin OH, Liu SH, Jun AS, Broman KW, Stark WJ, Vito EC, Narang AK, Thompson JM, Magovern M. Inheritance of a novel COL8A2 mutation defines a distinct early-onset subtype of fuchs corneal dystrophy. *Invest Ophthalmol Vis Sci* 2005;46(6):1934-1939

7 Chylack LT Jr, Wolfe JK, Singer DM, Leske MC, Bullimore MA, Bailey IL, Friend J, McCarthy D, Wu SY. The lens opacities classification system III. The longitudinal study of cataract study group. *Arch Ophthalmol* 1993; 111(6):831-836

8 Jurkunas U, Azar DT. Potential complications of ocular surgery in patients with coexistent keratoconus and Fuch's endothelial dystrophy. *Ophthalmology* 2006;113(12):2187-2197

9 Eghrari AO, Daoud YJ, Gottsch JD. Cataract surgery in Fuchs corneal dystrophy. *Curr Opin Ophthalmol* 2010;21(1):15-19

10 Barbour W, Saika S, Miyamoto T, Ohnishi Y. Biological compatibility of polymethyl methacrylate, hydrophilic acrylic and hydrophobic acrylic intraocular lenses. *Ophthalmic Res* 2005;37(5):255-261

11 Seitzman GD, Gottsch JD, Stark WJ. Cataract surgery in patients with Fuch's corneal dystrophy: expanding recommendations for cataract surgery without simultaneous keratoplasty. *Ophthalmology* 2005;112(3):441-446