Descemet membrane endothelial keratoplasty with a stromal rim in the treatment of posterior polymorphous corneal dystrophy

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A 20-year-old patient, diagnosed with posterior polymorphous corneal dystrophy, developed corneal edema for which he underwent Descemet membrane endothelial keratoplasty with a stromal rim (DMEK-S) in the right eye. No intra- or postoperative complications were noted. At the last follow-up 2 years and 9 months after the procedure, the best corrected visual acuity was 1.0 and endothelial cell density declined from 3533 cells/mm^2 to 1012 cells/mm^2. Despite the endothelial cell loss, DMEK-S appears to be a good alternative to other surgical techniques for the treatment of corneal endotheliopathies, and it may be of benefit to young patients.

Key words: Descemet membrane endothelial keratoplasty with a stromal rim, endothelial keratoplasty, posterior lamellar keratoplasty, posterior polymorphous corneal dystrophy

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Posterior polymorphous corneal dystrophy (PPCD) is an autosomal dominant disorder that affects primarily the endothelium and Descemet membrane (DM). Until now, a full thickness penetrating graft has been the gold standard for the treatment of endothelial failure in PPCD.[1,2]

Posterior lamellar keratoplasty is a relatively new surgical technique for the treatment of endotheliopathies. This approach offers several advantages compared to penetrating keratoplasty, such as a lack of suture-associated complications, less postoperative astigmatism and a quicker recovery.[3] Since the first introduction of this method in which the donor lamella consisted of the posterior stroma, DM and endothelium [Descemet stripping endothelial keratoplasty (DSEK)], many modifications have been reported.[3] The selective transplantation of lamella consisting of DM and the endothelium, referred to as DM endothelial keratoplasty (DMEK), is the most attractive approach.[4] Recently, a new technique of bare DM-endothelium transplantation – DMEK with a stromal rim (DMEK-S) – has been introduced.[5]

Case Report

A 13-year-old asymptomatic male was diagnosed with PPCD. At that age, his unaided visual acuity was 1.0 bilaterally. The posterior corneal surface was uneven, and there were opacities of DM in the corneal periphery. Changes of the posterior corneal surface were documented with a specular microscope (Noncon Robo Pachy SP-9000; Konan Medical Inc, Tokyo, Japan) [Fig. 1]. By the age of 20 years, both his corneas had become edematous, and his best corrected visual acuity was reduced to 0.4 in both eyes. After signing an informed consent, the patient underwent DMEK-S in the right eye. The study adhered to the tenets of the Declaration of Helsinki.

The donor button was prepared from a cornea stored under hypothermic conditions at 4°C in Eusol-C (Alchimia s.r.l, Padova, Italy). Donor age was 69 years, and the button consisted of a 6.0-mm diameter bare DM-endothelium lamella surrounded by a 1.0-mm wide posterior stromal rim; the endothelial cell density was 3533 cells/mm^2. The exact surgical technique, including the preparation of the donor button, has been reported elsewhere.[5] No complications were encountered [Fig. 2].

The clinical outcome was satisfactory. At the last follow-up 2 years and 9 months after the surgery, the patient had no complaints, and the cornea was clear with an unaided visual acuity of 0.5 and best corrected visual acuity 1.0 with −1.25 diopter sphere. The central endothelial cell density measured

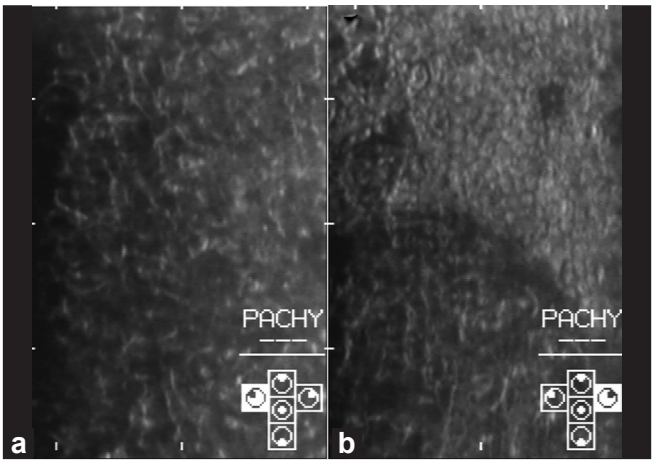

Figure 1: Specular microscopy images of the case with posterior polymorphous corneal dystrophy taken at the age of 13 years; right eye (a), left eye (b)

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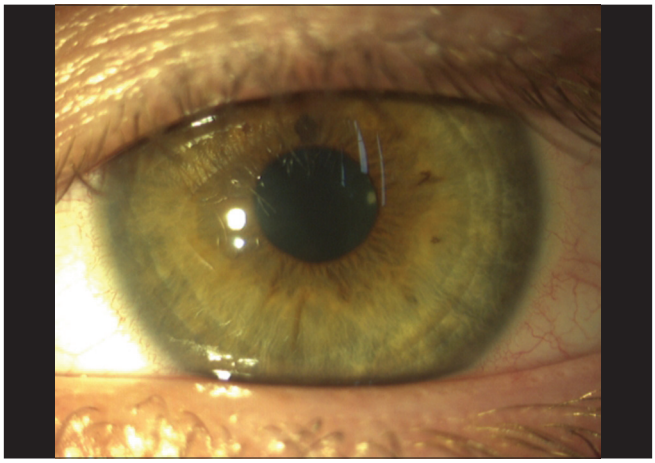

Figure 2: Right eye 1 month after DMEK-S (note the scleral rim of the lamella)

with a Topcon SP3000p noncontact autofocus specular microscope (Topcon Corp, Tokyo, Japan) 11 months after the surgery was 1089 cells/mm^2 and it was 1012 cells/mm^2 at the last follow-up nearly 3 years after the procedure.

Discussion

This is the first report of a posterior lamellar keratoplasty performed in a cornea with PPCD, using lamellae with bare DM and endothelium in the optical axis.[5] Although the necessity for keratoplasty differs in various cohorts of PPCD patients (it can be as high as 40%)[1,6] and a large majority of affected individuals are asymptomatic till an older age, many PPCD patients are younger at the time of grafting[1,2] than patients with late-onset Fuchs endothelial corneal dystrophy which is also treated with DMEK and DMEK-S.[4,5] Therefore, all possible benefits as well as disadvantages of various keratoplasty surgical techniques need to be very carefully evaluated.

The reason why we chose DMEK-S and not penetrating keratoplasty was that we considered it important to offer a young patient with an endothelial dystrophy the well-known benefits of posterior lamellar keratoplasty.[3] Compared to DSEK, the lamellae used in DMEK-S are thinner, allowing for an almost normal anatomic structure in the visual axis. Although the technique requires the acquisition of additional surgical skills, the scleral rim makes the manipulation with the lamellae easier than in DMEK, in our opinion. In addition, no instruments other than those used in routine penetrating keratoplasty and cataract surgery are needed in DMEK-S, and the surgery can be performed repeatedly.[5] The clear limitation of DMEK-S is the endothelial cell loss, in the case presented here 69% at 11 months, and 71% at 2 years and 9 months after the surgery, which implies that the technique needs to be further improved.

In the PPCD patient reported herein, DMEK-S led to the rapid recovery of vision without postoperative complications; however, the long-term survival of these grafts in the treatment of various conditions needs to be evaluated.

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