Descemet stripping with endothelial keratoplasty (DSEK) has gained popularity for the treatment of endothelial disorders. In this technique, the Descemet membrane and endothelium are stripped, and a donor posterior lamellar disc is inserted and allowed to unfold flush to the bare stromal surface. A crucial step to promote disc attachment is filling of the anterior chamber with air to push the donor tissue up against the recipient cornea. In aphakic patients, maintenance of air in the anterior chamber is a challenge. In this small case series, we provide the results of DSEK in 3 aphakic patients.

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Report of Cases. All the patients were part of a prospective clinical study of the safety and efficacy of DSEK in patients with endothelial diseases. Informed consent was obtained in accordance with the institutional review board of Bascom Palmer Eye Institute (Miami, Florida) and the Declaration of Helsinki.

Case 1. A 75-year-old man was first seen with a history of aphakic bullous keratopathy in his left eye. He had undergone extracapsular cataract extraction in 1973. In his left eye, the best-corrected visual acuity was counting fingers, and the intraocular pressure (IOP) was 18 mm Hg. At the end of uneventful DSEK, the anterior chamber was completely filled with filtered air. The patient was left face up for 1 hour, and afterward 50% of the air was evacuated. One drop of 1% cyclopentolate was instilled to prevent pupillary block. On the first postoperative day, the disc was attached, 40% air was present, and the IOP was 16 mm Hg. Blood was present at the interface peripherally, secondary to the interface blood (Figure 1B and C). Corneal edema progressively resolved, and at postoperative month 1 the visual acuity was 20/100.

Figure 1. Case 1. A, Slitlamp photograph shows successful button apposition 1 week after Descemet stripping with endothelial keratoplasty (DSEK). Arrows indicate blood at the interface that is not in the visual axis. B, Optical coherence tomograph of the cornea after DSEK. Note the meniscus-shaped DSEK button. Arrow indicates the area of increased signal intensity, probably representing interface blood. C, Closer view of the optical coherence tomograph of the cornea after DSEK. Note that the button is well apposed. Arrow indicates the area of interface blood.
Case 2. An 85-year-old woman was first seen with a history of pseudophakic bullous keratopathy in her left eye. She had undergone cataract surgery with scleral fixation of an intraocular lens (IOL) in 1992. In the left eye, the best-corrected visual acuity was hand motions, and the IOP was 21 mm Hg. Posterior pole ultrasonographic findings were normal. Intraoperatively, the IOL dislocated posteriorly, and a pars plana vitrectomy with lens retrieval was performed through an enlarged scleral tunnel incision. Retinal examination revealed an attached retina. She was left aphakic. The donor corneal disc was attached under an anterior chamber full of air. One drop of 1% cyclopentolate was applied, the patient was left face up for 1 hour, and 70% air was left. On the first postoperative day, the air bubble and the corneal disc were absent. Ultrasonographic results showed the donor corneal disc in the inferior anterior retina (Figure 2). After penetrating keratoplasty, a total retinal detachment developed, and she had no light perception.

Case 3. A 58-year-old man was first seen with a history of recurrent retinal detachments in his right eye due to high myopia. He had undergone extracapsular cataract extraction, vitrectomy, and silicone oil placement in the 1980s. In his right eye, the best-corrected visual acuity was 20/100. Examination revealed bullous keratopathy with silicone oil in the anterior chamber and vitreous and a large iridectomy inferiorty (Figure 3A), and the IOP was 12 mm Hg. Fundus examination revealed a normal optic nerve with an attached retina. The patient underwent DSEK with silicone oil removal. Intraoperatively, the air bubble could not be maintained in the anterior chamber, and sulfur hexafluoride (SF6) was injected into the anterior chamber to 100% fill and was later evacuated to 70% fill. The gas bubble remained, with good apposition of the donor corneal disc postoperatively. In the first postoperative month, his best-corrected visual acuity was 20/40, and the cornea was clear (Figure 3A). Subsequently, recurrent retinal detachments developed, but the donor corneal disc remained attached with corneal deturgescence (Figure 3B and C).

Comment. Descemet stripping with endothelial keratoplasty preserves the cornea’s biomechanical properties and may result in more rapid visual recovery than penetrating keratoplasty. The safety and efficacy of DSEK in aphakic eyes still needs to be determined. Introduction of air into the anterior chamber promotes attachment of the donor corneal disc to the recipient cornea. Maintenance of air is difficult in aphakic eyes in which there is no capsular bag to isolate the anterior chamber.

Price and Price described 2 eyes with aphakic bullous keratopathy before DSEK. A secondary IOL was implanted with DSEK in one eye, and a secondary IOL was implanted 4 months after DSEK in the other eye. A disc detachment found in one eye was successfully repositioned. In the present study, we described 3 aphakic patients who underwent DSEK with varied results. In the first patient, air was maintained postoperatively, and a favorable outcome was found. In the second patient, a serious complication occurred in which the donor disc displaced posteriorly. The intraoperative enlargement of the incision may have allowed a potential route for air to escape from the anterior chamber. In the third patient, owing to the large iridectomy, the air bubble disappeared behind the pupil. A longer-lasting gas ($SF_6$) with higher buoyancy than air was used to fill the anterior chamber, and good apposition of the donor disc was found postoperatively.

The main event leading to a favorable outcome in these cases may be the ability to maintain air in the anterior chamber in the first postoperative hours in addition to strict face-up positioning. The use of long-lasting, higher-buoyancy gases could be a possible solution for these patients. Ellis and Cohen showed the use of $SF_6$ for the treatment of Descemet membrane detachments after cataract surgery. Recently, some researchers have suggested using a suture to pull the disc into the eye and secure the disc to the recipient cornea. This technique may be of particular use in aphakic eyes, in which maintenance of an air bubble is more difficult.

In conclusion, owing to the small sample size and the retrospective nature of the study, we cannot conclude a lower rate of success of DSEK in aphakic patients. The possibility of a lower success rate of DSEK in aphakic eyes compared with pseudophakic or phakic eyes is an issue.
that needs further study. The potential for posterior dislocation of the donor corneal disc in aphakic eyes must be taken into consideration. Longer-lasting, higher-buoyancy gases (such as SF₆) could be used as alternatives to air, but the possible toxic effect to endothelial cells should be elucidated.

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Cherry Red Spot in Sialidosis (Mucolipidosis Type I)

The differential diagnosis of a cherry red spot in the macula includes central retinal artery occlusion and metabolic storage diseases such as Tay-Sachs disease, Sandhoff disease, Niemann-Pick disease, Fabry disease, Gaucher disease, and sialidosis. We report a case of an adolescent who, at a routine ophthalmic examination, was found to have a cherry red spot in the maculae of both eyes. Laboratory investigation results showed that the patient had mucolipidosis type I, which is a rare lysosomal storage disease with clinical and histologic findings similar to the mucopolysaccharidoses and the sphingolipidoses.

Report of a Case. A 14-year-old white boy complained of difficulty seeing the blackboard at school. A screening eye examination found decreased distance vision in both eyes. He was of normal intelligence and his medical history was significant only for scoliosis and seasonal allergies. Visual acuity corrected to 20/20 OU. Results of a dilated ophthalmoscopic examination revealed a cherry red spot in both maculae (Figure). Fluorescein angiography demonstrated hypofluorescence around the foveal area in the midvenous phase (eFigure 1, available online at http://www.archophthalmol.com). There was relative hyperfluorescence in the foveal area, but this was believed to represent a normal fluorescein pattern, rather than pigment epithelial disease. Optical coherence tomography showed increased reflectivity of the inner retinal layers, corresponding to the hypofluorescence on the fluorescein angiogram (eFigure 2). Because