Cataract Surgery with Foldable Single Piece IOLs in Congenital Cataract-Microcornea Syndrome

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ABSTRACT

Objective: To present the visual outcomes and intraocular lens (IOLs) stability after cataract surgery with foldable single piece IOLs in a patient with congenital cataract-microcornea syndrome (CCMC).

Case presentation: A 28-year-old woman presented with bilateral microcornea and posterior polar cataract. Her uncorrected (UDVA) and corrected (CDVA) distance visual acuities were 20/100, 20/160 and 20/80, 20/80, respectively. The horizontal cornea diameter was 9.0 mm in both eyes. Anterior chamber angle assessment (ACAA) with Scheie classification showed grade 3 on the superior, inferior, and temporal site and grade 2 on the nasal site in both eyes. On ultrasound biomicroscopy (UBM), the anterior chamber depths were 2.35 mm. OD and 2.41 mm. OS. The axial lengths were 23.28 mm., OD and 22.50 mm. OS with the A-scan ultrasound biometry. The IOLs power calculation with SRK/T formulas was +25.00 diopter OD and +22.50 diopter OS (A-constant 118.4). Phacoemulsification was performed on both eyes. After lens aspiration, ruptured posterior capsule, a common complication occurred. Then anterior vitrectomy was performed. A foldable single piece IOLs without haptic trimming was implanted in the sulcus in each eye. CDVA was 20/63 equally in both eyes. The intraocular pressure was 12 and 14 mmHg. ACAA showed grade 2 and no pigment dispersion. The IOLs had no tilt or decentration. UBM showed the IOLs were in the proper position and were not rubbing the iris.

Conclusion: The foldable single piece IOLs without haptic trimming in the sulcus were safely implanted in both eyes with microcornea in CCMC.

Keywords: Microcornea; posterior polar cataract; intraocular lens (Siriraj Med J 2017;69: 388-390)

INTRODUCTION

Congenital cataract-microcornea syndrome (CCMC) is the combined association of developmental cataract, microcornea and other rare ocular manifestations, including myopia, iris coloboma, sclerocornea and Peters anomaly without any other systemic anomaly. It is either an autosomal dominant or recessive inherited disease.[1-3] Bilateral posterior polar cataract is the most common cataract in CCMC. Microcornea is an abnormally small cornea with a horizontal diameter of less than 10 mm. Cataract surgery in eyes with CCMC is challenging because of issues related to the choice of anesthesia, surgical technique, decision to implant an intraocular lens (IOLs), intraoperative complications and limited postoperative outcome.[4] Due to anatomical abnormality, these eyes have a greater incidence of posterior capsular rupture, and they require a careful preoperative management plan. The authors described our experience with single piece posterior chamber IOLs in both eyes with CCMC.

CASE PRESENTATION

A 28-year-old woman presented with CCMC. She had bilateral microcornea and posterior polar cataract. (Fig 1) Her uncorrected (UDVA) and corrected (CDVA)

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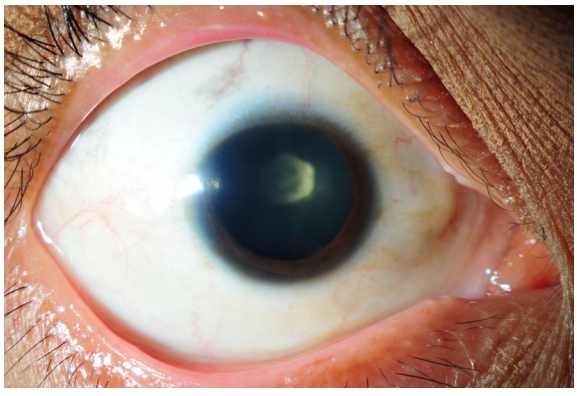

Fig 1. Microcornea and posterior polar cataract in congenital cataract-microcornea syndrome.

distance visual acuities using the LogMAR chart were 20/100, 20/160 and 20/80, 20/80, respectively. The horizontal cornea diameter was 9.0 mm in both eyes. Anterior chamber angle assessment (ACAA) with Scheie classification showed grade 3 on the superior, inferior, and temporal site and grade 2 on the nasal site in both eyes. The axial lengths were 23.28 mm., OD and 22.50 mm., OS from the A-scan ultrasound biometry. Keratometry was 41.82 D at /43.95 D, K1: 41.82 D × 174° (corneal curvature), K2: 43.95 D × 84°. The IOLs power calculation from the SRK/T formulas was +25.00 diopter OD and +22.50 diopter OS (A-constant 118.4). The anterior chamber depth was 2.35 mm., OD and 2.41 mm., OS from UBM. (Fig 2) She did not have nystagmus.

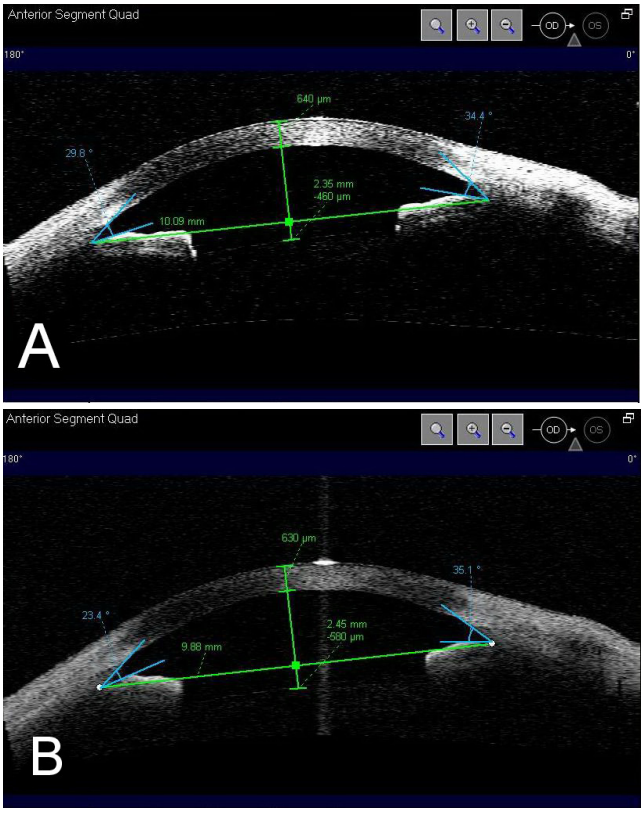

Fig 2. UBM showed the anterior chamber depth of the right eye before (A) and after (B) cataract surgery.

Cataract surgery was performed on the left eye under topical anesthesia. Lens aspiration was carried out after hydrodelineation without hydrodissection or rotation. After the epinucleus was aspirated, we found that the posterior capsule was torn and anterior vitrectomy was performed. A foldable single piece IOLs, modified L haptic design, without haptic trimming was implanted in the sulcus and captured the optic through the anterior capsulorhexis. The power of IOLs was adjusted according to the sulcus placement.

On the day after the surgery, the CDVA was 20/63 and there was no corneal edema. The patient was prescribed a combination of steroid and antibiotic eye drops for a month. At a two-year follow-up, the UCVA and CDVA of the left eye was 20/63. The intraocular pressure was 12 mmHg. ACAA showed grade 2 and there was no pigment dispersion. The IOLs had no tilt or decentration. UBM showed the IOLs was in proper position and did not rub the iris. The anterior chamber depth was 2.48 mm. The same surgery was performed on her right eye two years after the left eye. At a six-month follow up, the UCDA and CDVA of the right eye was 20/63 with normal IOP, normal ACAA, and well positioned IOLs. The anterior chamber depth was 2.45 mm. from UBM. (Fig 2)

DISCUSSION

Posterior polar cataract is a special challenge because it has a tendency for posterior capsule rupture and vitreous loss during cataract surgery.[5] Posterior capsular rupture is related with poor visual outcome.[6] Cortical cleaving hydrodissection can cause posterior capsule rupture, so it is a contraindication in posterior polar cataract surgery.[5] Hydrodelineation is recommend in these cases. Low aspiration flow rate, vacuum and bottle height will prevent posterior capsule stretching and reduces turbulence in the eye.[5] Implantation of an appropriate IOLs should be considered. It might be either anterior chamber IOLs or posterior chamber IOLs in the sulcus if there is adequate zonular support.[7] An acrylic 3-piece IOLs in the sulcus with optic capture will preserve the physiologic plane of the IOLs, reduce IOLs decentration or posterior iris chafing from the optic edge and prevent vitreous prolapse.[7] Iris-sutured IOLs, Glued-IOLs and trans-scleral IOLs have also been described.[7] When there exists a predisposition to posterior capsular rupture, it is important to carefully plan the operation and educate the patient with respect to the risk of drop nucleus, inability to implant the IOLs, and other potential complications.

In microcornea, cataract surgery carries the risk of complications such as corneal edema, posterior capsular

rupture, and unexpected aphakia.[4] There is only a small space in which to implant the IOLs. Haptic trimmed IOLs were mostly chosen to reduce complications such as glaucoma. Some reported studies indicated that scleral tunnel phacoemulsification reduced corneal edema in these compromised eyes.[4] Anterior chamber IOLs surgery is not recommended because of the risk for endothelial damage caused by a shallow anterior chamber and a difficult IOLs sizing in the anterior chamber. Recent studies have shown that the Glued-IOL implantation is effective because it has less pseudophacodonesis, good IOLs stability, less sizing difficulty and no polypropylene (Prolene) suture-related complications.[8] Haptic trimming has helped in the proper positioning of the IOLs, and the trimmed edge might strengthen adhesion along with glue.

In the present case, the authors decided to implant a foldable single piece IOLs without haptic trimming in the sulcus with optic capture, beacause a three piece IOLs may prove to be too large in the sulcus of patients with microcornea, and a haptic trimming IOLs might be unstable in the sulcus. Sharp trimming edges might also cause injury to the iris in the patient and could induce microhyphema, iris transillumination, pigment dispersion syndrome, pigmentary glaucoma, cystoid macular edema and uveitis-glaucoma-hyphema (UGH) syndrome.[9,10] One piece IOLs might decenter because the haptic loop-to-loop dimension of IOLs is only 13.0 mm.[11] The current authors considered it might fit in this microcorrnea patient with a small corneal diameter. The authors also considered that the haptic might rub on the iris and induced complications. However, this patient had ACD and axial lengths in the normal range,[11] so there was enough space for the haptic in the sulcus. Finally, the left eye did not develop secondary glaucoma, pigment dispersion syndrome, iris transillumination or any complications after a two year follow up. The ACD was slightly decreased and the IOLs was stable. So the authors decided to operate on the right eye and the results were the same after a six month follow up.

CONCLUSION

The foldable single piece IOLs in the sulcus, without haptic trimming, was safely implanted in both eyes with microcornea.

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