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Case report

Long-term visual and ocular health outcomes of 2 sets of bilaterally aphakic siblings utilizing contact lens correction

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ABSTRACT

We report the long-term clinical courses of 8 aphakic eyes of 2 sets of siblings who used contact lenses for both refractive correction and amblyopia treatment following neonatal cataract extraction. Early cataract removal, aggressive contact lens use, and robust professional supervision seem to have substantially contributed to visual success in our four patients. All eyes did well visually with contact lenses, all developing acuities close to 20/20 despite contact lens and non-contact lens related complications that were managed. Complications of most concern were corneal neovascularization and glaucoma. We believe this to be the first case series documenting consistent long-term visual and ocular health outcomes of sets of bilaterally aphakic siblings optically treated with contact lenses.

1. Introduction

Congenital cataract is not a common finding at birth but it is one of the leading causes of blindness in children worldwide [1,2]. Approximately 1.5 in 10,000 infants have cataracts at birth in the United States [1]. Visual prognosis is greatly improved in these children with surgical cataract extraction [3,4] prior to 8 weeks of age [1,5–7]. Factors that have shown to play a significant role in visual outcome of children with congenital cataracts include: the type of cataract, appropriate refractive correction immediately following the cataract surgery, and initiation of aggressive amblyopic treatment [1,4–8]. Bilateral aphakic patients have been generally shown to achieve better visual acuities than unilateral aphakic patients [6,8]; unilateral aphakia more often leads to greater degree of amblyopia [1,4,5]. We are unaware of any literature discussions of bilaterally aphakic siblings.

Visual outcomes of cataract surgery often depend on postoperative complications. The most common and the most sight threatening complication of congenital cataract surgery is glaucoma, with a reported incidence of between 15% and 45% [9–11]. Studies show that there is a marked increase in the risk of secondary glaucoma if the cataract is removed within the first month of life [1,3,5–7,9,12]. Haargaard et al. also found the rate of glaucoma to be 7.85-fold higher in children who were less than 9 months of age at surgery compared to those that were older [3]. Besides the age at surgery, another study found the risk of secondary glaucoma to be greater if aphakia is bilateral but the reason remains unclear [12]. Measuring and monitoring intraocular pressures in young children is challenging however, leading clinicians to rely on other signs to make the diagnosis of secondary glaucoma: an excessive drop in aphakic refractive error, enlarged corneal diameters, and optic nerve cupping [13,14].

Contact lenses (CLs) have been the standard of care in the optical correction of aphakia [1,5,15], especially in children younger than 2 years of age [2], for about 40 years. A wide variety of highly oxygen permeable soft silicone hydrogel and rigid contact lens designs are available today, replacing older non-permeable rigid and low permeability traditional hydrogel materials. Many of the new materials, rigid or soft, are designed to allow sufficient oxygen to reach the cornea during daily wear, even allowing for centrally very thick aphakic lens optics, so that the CL driven complication of hypoxia should no longer be a concern. In addition, the newer materials may be optically more stable compared to the older hydrogel lens material that are prone to dehydration, which could cause power reduction on the eye [16].

Intraocular lens (IOL) technology and microsurgical techniques have improved as well during the last several decades and as a result, the use of intraocular lens implantation for the optical correction of adult aphakia has become dominant [1,2,5,15]. The use

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of IOL in infants is still controversial, however, due to known associated postoperative complications.

Many studies have previously documented the long-term results of using CLs for infantile aphakia, especially unilateral aphakia [1,2,5,15,17]. Approximately 65% of infantile cataracts are bilateral however [1], and yet, there are few reports on visual prognosis and ocular complications of bilaterally aphakic children managed by contact lenses. We believe this to be the first case series documenting long-term visual and ocular health outcomes of sets of bilaterally aphakic children optically treated with contact lenses.

2. Case series

Our case series presents four patients: two sets of siblings with bilateral congenital aphakia who were all followed aggressively for many years by both a CL practitioner and a pediatric ophthalmologist, beginning with cataract removal as infants.

Note: All four patients were initially overcorrected by +3 to +4 diopters for near vision until they began to walk, at which point they were given contact lenses with optical powers for appropriate distance correction, as defined by retinoscopy, and a pair of bifocal spectacles for safety and near vision to be worn over contact lenses.

2.1. Case 1

In January 1982, a 12-week old baby boy presented to the Contact Lens Service at the Jules Stein Eye Institute. He had been diagnosed with bilateral congenital cataracts soon after birth and underwent cataract extraction in each eye on two separate days at 10 weeks of age. At the initial visit, retinoscopy findings were +30.00DS in the right eye and +24.50DS in the left eye. An ultrasound was performed to measure the axial length of both eyes to help predict the needed powers of the CLs. Then a diagnostic hard CL was placed on each eye sequentially to allow estimation of corneal curvatures. Based on approximated corneal curvatures, an appropriate initial diagnostic soft CL was selected and placed on each eye sequentially for over-retinoscopy. From this evaluation, a pair of custom soft CLs was ordered at the end of the initial visit. The patient returned to clinic ten days later. His first pair of soft CLs were evaluated and dispensed and the parents were trained for insertion and removal. The patient returned to clinic every 2–4 weeks during the first year of CL wear for assessment of tolerance, mechanical fit, and optical powers of his CLs and for replacement of lost lenses. Each eye went through several modifications of both soft lens base curves and powers during the first year: 6 in the right eye and 14 in the left eye. The CL modifications became less frequent beyond 15 months of age so he was evaluated every 2–4 months thereafter.

At the age of 8, he achieved 20/40 each eye on the Snellen chart with soft CLs. At this time, he was refitted into rigid gas permeable (GP) lenses due to residual astigmatism observed with the soft CLs. Three months after the refit, his visions were noted to have improved to 20/30 each eye. At 9 years of age, his visual acuities with GP lenses achieved 20/20 each eye.

Throughout the 25 years of CL care at the Contact Lens Service, the patient did not develop any contact lens related complications. However, he developed strabismus and amblyopia in his left eye in 1984 and his pediatric ophthalmologist promptly initiated amblyopia therapy. Glaucoma was diagnosed in the left eye in 1994 and then in his right eye in 2001. Glaucoma medications were prescribed following each diagnosis, eventually: topical latanoprost q.h.s. and dorzolamide b.i.d. in the right eye and betaxolol b.i.d., brimonidine b.i.d. and dorzolamide t.i.d. in the left eye. A co-managing ophthalmologist has been closely monitoring his intraocular pressures which have remained stable and well-controlled by maximum medical therapy over the years.

The patient was last seen in the Contact Lens Service in 2006 at age 24. At his last visit, his visual acuities were 20/20 in each eye with GPs. With a manifest refraction of +15.00DS, he achieved 20/20 right eye and 20/25 left eye. Keratometry readings were 40.75DS with trace distortion right eye and 40.75/40.00 @ 154 with trace distortion left eye.

2.2. Case 2

A 6-week old baby girl, the younger sister of the patient above, presented to the Contact Lens Service at the Jules Stein Eye Institute in December 1983 after having undergone cataract surgery for bilateral congenital cataracts at the age of 4 weeks in two separate surgeries. At this visit, she was fitted into regular custom soft CLs based on her retinoscopy findings of +35.00DS in both eyes and estimated corneal curvatures (methodology similar to Case 1). She was initially followed every 2–4 weeks for her CL care.

She only needed a few base curve and power modifications during the first year of CL wear. After the first year, her CL parameters stabilized and subsequently managed for CL care every 2–4 months. In 1990, after her brother had been successfully refitted into rigid GP lenses, she also returned with the request for a similar GP refit. After the refit, she achieved 20/30 acuity in each eye. In 1998, her CL corrected visual acuities were noted to have improved to 20/20 each eye.

She did not develop any contact lens driven complications throughout the 27 years of CL care at Jules Stein Eye Institute. However, she was diagnosed with disassociated vertical deviation in 1988 and ocular hypertension in 1999 by her pediatric ophthalmologist.

Her most recent contact lens evaluation was performed in August 2010 at age 26. On this visit, her acuities with custom GP lenses were 20/25+ right eye and 20/20 left eye. Manifest refraction was +16.75DS with 20/25+ acuity right eye and +17.50DS with 20/20 acuity left eye. Keratometry readings were 43.37DS with 1+ distortion right eye and 43.12/42.50 @ 180 with trace distortion left eye. Her intraocular pressures were stable without any intraocular pressure lowering medications and are routinely monitored by the co-managing ophthalmologist.

2.3. Case 3

In 1997, an 11-day old boy with a history of congenital bilateral cataracts and cataract extraction in the right eye at 5 days of age and left eye at 10 days of age, presented to the Contact Lens Service at the Jules Stein Eye Institute. At this visit, retinoscopy findings were +19.00DS right eye and +21.00DS left eye. A diagnostic soft CL was placed on each eye to determine initial optical powers and base curves needed by performing over-retinoscopy followed by a slit lamp examination with the use of high molecular Fluoresoft™.

Despite strong professional recommendation to use custom soft CLs for only daily wear, the parents insisted on soft CLs for extended wear because they were worried about their son taking frequent naps. The patient returned to the office every 1–2 weeks during the first 3 months of his CL wear to monitor the health of the eyes, to make any necessary CL modifications to keep up with his rapidly growing eyes and to replace torn or lost lenses. Our patient's parents were frequently advised to allow a refitting into daily-wear soft lenses for ocular health and for broader range of lens parameters, especially optical power, they offer.

At 9 months of age, the patient was refitted into the daily-wear soft lenses. However, his mother reported he continued to occasionally nap in his CLs and eventually circumferential corneal neovascularization was noted in both eyes (in 2000). He was immediately refitted into rigid GP lenses but tolerance was poor and his conjunctivae were constantly inflamed with GP CL wear despite