“My Most Challenging Contact Lens Case” - Contact Lens and Refractive Management of Spherophakia with Glaucoma

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Abstract: A 10-year-old patient with spherophakia was referred to our clinic for a contact lens fitting. The patient was treated with bilateral twice-a-day atropine and timolol therapy. After considering several options, she was fitted with plano prosthetic iris soft lenses piggybacked with gas permeable lenses, which provided correction for her high myopia. This modality, coupled with near vision spectacles, provided the patient with good distance and near acuity and eliminated photophobia caused by atropine therapy.

Keywords: spherophakia, lenticular myopia, pupillary block glaucoma, atropine therapy, prosthetic iris contact lens, piggyback contact lenses
Background

Spherophakia (aka microspherophakia) is a rare condition wherein the crystalline lens assumes a shape more spherical than the typical anatomical lens, having a smaller equatorial diameter and a larger antero-posterior diameter. [1] Progressive lenticular myopia, caused by the highly curved anterior lens surface and anterior displacement of the lens, is a refractive finding in all spherophakic patients. Lenticular myopia and spherophakia are progressive between the 8th and 15th years of life; significant refractive and anatomical changes should be expected during this period.[2] Additional ocular findings of spherophakic eyes are ectopia lentis, hypoplastic ciliary body, nanophthalmos and secondary glaucoma.[3,4] The mechanism for the development of glaucoma in spherophakia is pupillary block. It is theorized that spherophakia develops as a result of a lack of support from the zonular-ciliary body complex. The presence of a hypoplastic ciliary body and long zonular fibers found in these patients suggests that the fetal lens, which is naturally spherical, has not been reshaped by the forces of the ciliary body and zonules. The mean antero-posterior diameter of a spherophakic lens is 1mm larger than a normal age-matched lens. This, in combination with loose zonule fibers, predisposes these patients to pupillary block and angle closure.[5]

We present the case of a young female patient diagnosed with spherophakia and treated with peripheral iridotomies, timolol and atropine. Due to her high myopia, increasing refractive error, large pupils and lack of accommodation, the patient required specialty refractive care. By combining soft prosthetic lenses, rigid gas permeable (GP) lenses and a near vision spectacle prescription, the patient achieved good vision and visual comfort.
Case Report

A 10-year-old Hispanic female was referred to our contact lens clinic for refractive management of her high myopia and the side effects of her glaucoma treatment. The patient was of small to average build, normal to high mental capacity, and normal behavior. Several systemic conditions have been associated with spherophakia, including Marfan’s, Marchesani’s, Alport’s and Klinefelter’s Syndromes, none of which were present in our patient. [6] The patient had been diagnosed with spherophakia and was using Atropine sulfate ophthalmic solution 1% (Falcon Pharmaceuticals, Fort Worth, TX) BID OU and Timolol maleate ophthalmic solution 0.5% (Ista Pharmaceuticals, Irvine, CA) BID OU, which, combined with bilateral laser peripheral iridotomies, successfully controlled her pupillary block glaucoma. On examination at referral, the patient’s best-corrected Snellen acuity was 20/40 OD with -20.25-0.50 x 005 and 20/40 OS with -17.50 DS (12mm VD). The patient entered wearing single vision high index spectacle lenses. Several initial concerns were elicited from the patient and from her parents. The patient’s mother was concerned about the fact that the spectacle prescription had been increasing every 3-5 months for the past few years. She was also concerned that her child seemed to be uncomfortable outdoors and in bright lights, and was unable to wear sunglasses with her spectacle correction. The child mentioned that she had some difficulty at school, as near work and reading were often blurry. Slit-lamp evaluation demonstrated normal adnexa, lids, conjunctiva and cornea OU, the anterior chambers were shallow, and the iridies were flat with patent laser peripheral iridotomies. The physiologic lenses were visible, secondary to well-dilated pupils and were small, with zonules clearly visible around the lens and with the anterior portion of the lens extending through the pupillary
zone anteriorly. The pupils were pharmacologically dilated (9.5-10.0mm), round, and non-reactive to light.

At the first visit, the patient was refracted, and after education on more effective visual use, was given a single vision spectacle prescription to be filled in a polycarbonate material. The patient was also fit with spherical gas permeable contact lenses. Her keratometry readings, as measured by topography were OD 44.4/42.8@010 and OS 44.7/42.8@163. Trial lenses were fit with lid attachment, and provided good stability, centration and movement as well as an aligned central fluorescein pattern and an average edge lift. The initial lens parameters were ordered (Table 1) from X-Cel Contact Lens Company (Walman Optical, Minneapolis, MN) in Boston XO material.

Table 1: GP Lens Parameters

<table>
<thead>
<tr>
<th>Eye</th>
<th>Lens Design</th>
<th>Base Curve</th>
<th>OAD</th>
<th>OZD</th>
<th>Power</th>
</tr>
</thead>
<tbody>
<tr>
<td>OD</td>
<td>X-Cel Thin Custom</td>
<td>7.65</td>
<td>9.2</td>
<td>STD</td>
<td>-15.00</td>
</tr>
<tr>
<td>OS</td>
<td>X-Cel Thin Custom</td>
<td>7.60</td>
<td>9.2</td>
<td>STD</td>
<td>-15.25</td>
</tr>
</tbody>
</table>

The lenses were dispensed one week later with a good fit (central alignment, adequate edge lift, and lid attachment) and good initial comfort. The patient immediately enjoyed her improved acuity and field of view, and demonstrated 20/30 distance acuity OD, OS, and OU. However, the patient commented that she was able to see “something moving” when she blinked. A two-week trial with the GP lenses was agreed upon to assess the adaptation to the new lenses, increase wearing time, and evaluate the possibility of neural adjustment in perceiving the edge of the lens. Before leaving, the patient was given a prescription for plano/+2.50 progressive addition spectacle lenses for near vision use over her contact lenses. The patient, as well as her parents, performed lens care (with
UniquePH solution, Alcon, Fort Worth, TX), insertion and removal before leaving the office.

The patient returned for follow-up two weeks later, and reported that she was happy with her vision and had good comfort with the lenses. The patient was cleaning the lenses and doing much of the insertion and removal of the lenses on her own, under the supervision of her mother. The patient stated that she continued to notice the edges of the lens, specifically when reading and doing near work. Her mother again inquired about the patient’s indoor and outdoor light sensitivity. To address these problems the patient was fit with prosthetic iris lenses to be used in a piggyback system with the GP lenses. CooperVision (Fairport, NY) Standard Prosthetic DW lenses were fit (Table 2).

Table 2: Prosthetic Lens Parameters

<table>
<thead>
<tr>
<th>Eye</th>
<th>Lens Design</th>
<th>Base Curve</th>
<th>OAD</th>
<th>Pupil Diameter</th>
<th>Iris Color</th>
</tr>
</thead>
<tbody>
<tr>
<td>OU</td>
<td>Cooper Prosthetic DW</td>
<td>8.7</td>
<td>14.5</td>
<td>4.5</td>
<td>Dark Brown with Black Backing</td>
</tr>
</tbody>
</table>

The prosthetic iris lenses were ordered and dispensed to the patient. Upon insertion of the prosthetic lens/GP piggyback system, the patient immediately noted a resolution of peripheral lens awareness in her vision, as well as an improvement in visual comfort and photophobia, especially in bright light conditions. The prosthetic lenses were well fit, with good coverage, centration and movement. When the GP lenses were reinserted over the prosthetic lenses, they maintained good centration, lid attachment and movement, but demonstrated a slightly steep central fit upon fluorescein evaluation. The lenses were dispensed; the GP lenses will be reordered with a slightly flatter base curve to accommodate the piggyback fit. The patient was again instructed on the slightly
varied cleaning, insertion and removal necessary to care for soft lenses, and was prescribed Optifree Replenish (Alcon, Fort Worth, TX) for lens cleaning and storage. Although it is premature to judge the long-term success of this system, the patient’s initial response was very positive. She wears plano sun protective tinted lenses when outdoors and is no longer photophobic. She continues to be followed by the referring ophthalmologist for her glaucoma care, and is re-evaluated every 3 to 4 months in our contact lens clinic to determine whether contact lens changes are necessary. Her final acuity is 20/30 OD, OS, and OU at distance and near with her contact lenses and near vision spectacles.

Discussion

Fitting a high myope with contact lenses should be a mundane act in the day of a contact lens resident. However, contact lens management of a high myope with a rapidly changing refractive error, large pupils, no accommodation, photophobia, concomitant ocular disease, and a young age is bound to require some problem solving and unique ideas for management. The patient’s age, type of lens (or lenses) to be worn, long term effects of this mode of lens wear, cost of the chosen modality over time, special needs of a patient with a rare ocular condition, secondary concerns due to disease treatment, and additional time for discussions and explanations all combine to challenge the practitioner.

The management of glaucoma in patients with spherophakia is complicated and no definitive treatment is currently identified. Treatment with pharmacologic agents is often under-effective and has challenging side effects. Laser treatments are often only transiently effective, and surgical treatments carry a high risk of significant
complications. The pharmacologic management of spherophakia includes topical and/or oral aqueous suppressants, as well as mydriasis/cycloplegia. Cycloplegic agents relax the ciliary muscle causing a tightening of the lens zonules and posterior lens movement. [7] Mydriasis moves the iris to a peripheral location, and together with cycloplegia, this minimizes the potential of iris-lens contact and pupillary block. Side effects of this treatment include loss of accommodation, photophobia, and increased high order aberrations due to large pupil size. Peripheral iridotomy allows the free flow of aqueous from the posterior to the anterior chamber, even in the event of iris-lens contact. Possible complications of iridotomy include vitreous loss and zonular destruction, as the lens periphery is not present to protect these structures.[8] Surgical lensectomy becomes necessary for glaucoma control in some patients, and can often permanently control intraocular pressures. Lensectomy is a challenging procedure in spherophakes, as the lens is unstable, prone to dislocation, and can have a tough capsular bag. The anterior chamber is often very shallow, but anterior chamber intraocular lens (IOL) placement is almost always necessary if an IOL is to be used. Even after lensectomy, IOP is not always controlled, and additional surgical intervention is often needed to control IOP. Eyes undergoing pressure-lowering surgical procedures appear to have a propensity for developing anterior synechiae, which can block the aqueous filtering structures and negate the surgical effects. Specialists attempting large-scale surgical management of the condition through lensectomy suggest that a “satisfactory outcome appears to be rare”. [9]

Many ten-year-old patients are capable of wearing and caring for contact lenses independently. Our patient was excited to wear lenses and mature enough to do most of the handling and cleaning of the lenses on her own. Nevertheless, we also educated her
parents in the handling and cleaning of the lenses in order to assist her as she improved her skills. Many practitioners are uncomfortable fitting younger patients with lenses; however, others believe that a patient should not be excluded because of age, if they are motivated and mature enough to follow the instructions for lens wear and care. In this case, the time to instruct the patient in lens care, insertion and removal was similar to the time it takes other young people to complete this task. In fact, the act was much more challenging for her parents, who were not lens wearers.

Our patient initially entered wearing single vision spectacle lenses and complaining of near vision blur. While we initially questioned the decision to provide lenses of this type to a patient using atropine drops, we eventually chose to prescribe the same type of lens, but with additional education and instruction given to the patient. The options for spectacle lens wear without contact lenses were discussed, and ultimately, taking advantage of vertex distance changes with high powered lenses, we were able to teach the patient to adjust the position of her lenses to achieve clarity for near work, and prevent the need for a bifocal lens. Progressive lenses (PAL) with a +2.50 diopter add and plano distance power were prescribed for near vision use over contact lenses. To address the patient’s visual needs, we recommended 99% UV sun-protective lenses for use outdoors.

Our patient’s concerns for her near vision blur were addressed through education and redirection of single vision lens use, and through PAL use over her contact lenses. Her parents’ concerns over photophobia were addressed with the prosthetic iris contact lens, and with plano sun protective lenses. Previous to this experience, I had never worked with a patient with spherophakia, and as such, had limited knowledge of the
condition or its normal progression. After researching the topic, I learned why the patient was being managed pharmacologically instead of surgically, as well as the expected progression of myopia during the 8th to 15th years due to lenticular myopia, rather than axial changes. This information, as well as her mother’s reports of 3-5 month spectacle lens changes for the past several years, assisted us in choosing the best contact lens option for the patient.

Initially, we felt that a custom silicone hydrogel lens available in high powers (O2 Optix Custom, CIBA, Duluth, GA) would be ideal. Post-manufacture prosthetic iris prints are available through several companies (Adventure in Color, Golden, CO; Specialty Tint, Encantido, CA), but the cost is significant. The prospect of having to change the lenses frequently made this option prohibitive due to cost, and the time necessary to manufacture the lenses. Ultimately, we decided that the best option for the patient was to use a plano soft lens with a prosthetic iris, and to piggyback a GP lens over it. This alternative seemed to provide the greatest modification potential at the lowest future cost. The prosthetic lenses can be used for one year or more, and the GP lenses can be modified in office if small changes are needed, or can be reordered at a relatively low cost if significant changes are needed. The low Dk of available prosthetic lenses (Dk 8.4 – CooperVision Standard Prosthetic) is of concern for a young patient who will likely be wearing the lenses for many years. However, the need to provide the patient with comfortable vision outweighs the risk of corneal neovascularization. Emphasis on daily wear of lenses only, and proper care and handling can minimize potential problems. Our GP lens material was selected to have very high oxygen permeability (Dk 100 – Boston
XO) to minimize the contributory Dk reduction. Perhaps, over our patient’s period of lens wear, higher Dk materials will be available for prosthetic soft lenses.

When initially fitting the GP lens, it was expected that not only the optical zone, but also the overall diameter of the lens, would be much smaller than the patient’s pupil. The optics of such a system would be expected to cause visual disturbances with such a large pupil, but for purposes of lens adaptation, and for the possibility, while remote, that prosthetic lenses would not be necessary, we decided to do a short trial with GP lenses alone. As expected, the patient required the prosthetic lens to eliminate peripheral blur and photophobia.

While piggybacking rigid and soft lenses, it is not atypical to expect that the base curve of the GP lens may need to be modified to fit over the soft lens. In this case, the base curve needed to be flattened to maintain the same fit that it had before piggybacking. The necessary modification may change depending on the power and anterior curve of the soft lens.

Contact lens patients using prescription eye drops can present unique challenges. Drop insertion is not recommended during lens wear, and the side effects of prescription eye drops can often make lens wear less comfortable. Our patient was using two eye drops twice a day. Twice a day dosing can still be conducive to contact lens wear as drops are inserted prior to lens insertion, and after lens removal. In patients using eye drops in this way, it is important to inform them not only to wait 10 minutes between drop insertion, but also to wait 10-15 minutes after drop insertion before lens insertion to minimize absorption of the medication and preservatives by the lens. The minimal stinging and redness that can be associated with the insertion of Atropine or Istalol
appeared to be minimal or non-existent in our patient. However, the side effects of BID atropine are significant. Systemic side effects are possible from topical atropine, including tachycardia, confusion, flushing of face and skin, weakness and drowsiness. Our patient did not experience any of the possible systemic side effects of the medication, but did have a loss of accommodation and photophobia from its use.

The choices of solution for a patient using a piggyback lens system can be varied. Most patients will use two separate cleaning systems for their lenses, although the option for single solution cleaning is available with hydrogen peroxide solutions (Clear Care, CIBA Vision). Our patient trialed three rigid lens cleaning systems due to a stinging sensation from several of the solutions, ultimately she had the most comfort with Unique Ph, and chose to continue using it. Optifree Replenish was prescribed for soft lens disinfecting.

Patients referred for specialty contact lens fittings are often those with the most interesting cases and are the most rewarding at completion. However, it can be difficult to work with a patient when not all of the information about their ocular disease or eye care history is available. In this case, I had many questions about the medical/surgical management of the patient’s condition. With a moderate amount of research, I was able to further educate myself on the condition; I also had an opportunity to discuss the case with the referring ophthalmologist.

Conclusion

Spherophakia and its associated ocular complications can present a clinical challenge to the contact lens specialist because of the rapidly changing refractive status,
pharmacologically dilated pupils, and lack of physiological accommodation. Although imperfect, piggybacked soft prosthetic iris lenses, along with gas permeable lenses can provide excellent vision, physiologically acceptable ocular conditions, and overall patient satisfaction. However, it does make the clinician acutely aware of the complexity of the visual options, and the improvements that are still needed.

Figure 1: Spherophakic eye without prosthetic lens.

Figure 2: Spherophakic eye with prosthetic lens/ GP lens piggyback.
References


