Announcement

Winning Essay: Student Award in Clinical Ethics

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Andrea Meagher, OD, is the 2022 recipient of the Student Award in Clinical Ethics.
Her winning essay appears here.

Caring for Patients with Disabilities: Advocating for Those Who Cannot Advocate for Themselves

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As optometrists, most of us have at one time provided care to a patient with a developmental disability. Individuals with disabilities often experience discrimination in health care.[1] There is also a lack of optometric education and research in the area of patients with developmental disabilities.[2] This case study involving a patient with Down syndrome and keratoconus helps bring to light the positive impact optometrists can have on this population through not only ethical treatment and management, but also advocating for their well-being.

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Current clinical guidelines for management of keratoconus are based on whether patients are satisfied with their vision in glasses.[3] This begs the question: What if a patient is unable to express that satisfaction, or lack thereof? How can a clinician decide whether patients are satisfied with their current

vision if they cannot communicate such and have never had the opportunity to see the world clearly?

Patient Assessment

A 31-year-old female reported to the Cornea and Contact Lens department at Illinois Eye Institute for physician-directed follow-up for keratoconus. She had first been diagnosed with keratoconus in the Developmental Disabilities clinic at Illinois Eye Institute 2 weeks prior. She had undergone comprehensive eye exams at outside practices at least every 2 years since birth with normal results aside from refractive error. The patient's medical history was positive for Down syndrome and obesity. She was somewhat verbal, but spoke only Spanish. Her sister was present to translate during our exam. The patient had a positive family history of keratoconus. Her mother was the primary caretaker but her sister lived close by and was with her most days. Although the patient had been prescribed numerous pairs of glasses since age 5, she had never worn them successfully due to not liking the feeling of the frame on her face.

Her previous visual acuities were documented as "fixate and follow," but the patient was found to be very adept at Lea matching during her exam in the Cornea and Contact Lens department. She achieved a distance visual acuity of 20/200 in the right eye, 20/300 in the left eye, and 20/125 with both eyes through her glasses prescription. Her near acuity was measured as 20/40 in each eye, which indicated that she was likely not highly amblyopic. Pupil testing and extraocular motility testing were normal. Confrontation visual fields were unable to be tested due to patient understanding and poor fixation. Retinoscopy revealed a prescription similar to her previous glasses of -9.00 -2.00 x 028 in the right eye and -9.00 -4.50 x 160 in the left eye. A corneal tomography was obtained, although quality was slightly reduced due to poor fixation. It revealed keratometry readings of 58.40/60.60@099 in the right eye and 72.20/78.90@073 in the left eye with a minimum pachymetry thickness of 302 µm in the right eye and 155 µm in the left eye. The tomography also showed apical stromal corneal thinning consistent with keratoconus in both eyes. Slit lamp examination revealed mild central corneal scarring in the right eye and significant central corneal scarring in the left eye with a significant Munson sign upon down gaze in both eyes. The bulbar conjunctiva was slightly diffusely injected in both eyes. Upon instillation of sodium fluorescein there was a band of 1+ punctate epithelial erosion in the left eye with a question of incomplete lid closure/lagophthalmos. All other slit lamp findings were normal. The patient had her pupils dilated at her exam 2 weeks prior, and the posterior pole examination was found to be within normal limits with cup-to-disc ratios of 0.30/0.30 in both eyes.

Current clinical guidelines indicate that first-line treatment for keratoconus should be corneal crosslinking for those who are eligible.[3] Due to the patient's minimum corneal thickness of less than 400 µm and the central scarring present in both eyes, she was not an ideal candidate for corneal crosslinking.[3] The next line of treatment is specialty contact lenses.[3] Corneal transplant surgery is recommended only if contact lenses still cannot achieve adequate vision.[3] Despite more than 25 years of eye care and known high ametropia, the patient had never been offered the correction option of contact lenses to improve her vision and comfort. Had she not been developmentally delayed, one can suspect she would have been given the option of contact lenses much sooner.

Patience Leads to Progress

The American Optometric Association (AOA) Standards of Professional Conduct support that we have an ethical obligation to provide this patient with the option of contact lenses with the following statement: "Optometrists have a duty to inform patients or their legal guardian about the patient's health care and health care options."[4] This patient is not the only disabled patient who has been overlooked in this way. The prevalence of keratoconus in people with Down syndrome is higher than in the general population, but many patients go undiagnosed due to difficulty in providing care.[5] Sadly, patients with developmental disabilities are not being provided the same level of health care as other patients.[6] This is a violation of

AOA's Code of Ethics tenet "to strive to ensure that all persons have access to eye, vision, and general health care" ? not just those persons who are able to express their desire for functional vision.[7]

At this patient's first visit to the Cornea and Contact Lens department, we educated her accompanying guardians on her condition and management options. The guardians agreed to help the patient with application and removal of the lenses and to proceed with the contact lens fitting in hopes of achieving better vision and comfort than her habitual spectacles provided. We were able to apply a hybrid lens to her right eye (on the first attempt!) and the patient reported good overall comfort. We then attempted to apply a hybrid lens to the more severe left eye but encountered much greater difficulty. After repeated attempts, the patient became overwhelmed. We were unable to achieve an accurate visual acuity or fit evaluation of the lens, so it was removed and she was asked to return at the next available appointment to continue the fitting.

At her next fitting appointment 3 weeks later, the patient expressed her initial apprehension with the contact lens fitting process. With some time, reassurance and review of the process, she agreed to proceed with the fitting. Throughout this appointment she grew more and more comfortable with me and the lenses, and we were able to successfully apply and evaluate two hybrid lenses on the right eye and three hybrid lenses on the left eye. We adapted the fitting process to her specific needs: warming the preservative-free saline vials to reduce the blink reflex, providing a dental bib to catch any spills, and moving slowly with the utmost patience. The patient achieved best-corrected distance vision of 20/40 in the right eye and 20/30 in the left eye with over-refraction, a drastic improvement from 20/125 in her glasses! She expressed comfort with the lenses and amazement in her vision improvement that was truly heartwarming. Although we achieved an appropriate fit in the right eye, her left eye exhibited central touch with even the highest possible hybrid lens vault. For this reason, the patient was scheduled to return for an additional fitting appointment for scleral lenses. The hope is that she will be more cooperative with insertion of these larger gas permeable lenses after having had a positive experience with the hybrid lenses.

Fulfilling Our Responsibility as Optometrists

Down syndrome is not uncommon. It occurs in 1 of every 700 babies born in the United States.[8] Furthermore, the number of patients with Down syndrome is increasing due to increasing maternal age along with medical advances in technology resulting in better survival rates for these children.[9] It is our responsibility as optometrists to understand how to properly treat and manage the unique visual needs of this population. Although this patient was able to respond to Lea matching, most patients with Down syndrome respond best to Teller Acuity Cards.[9] In addition to keratoconus, blepharitis, premature cataract and strabismus are common in Down syndrome patients.[9]

Patients with Down syndrome and other disabilities who present with high ametropia, including high amounts of cylinder and a "scissor" reflex on retinoscopy, should be screened for keratoconus with corneal tomography. Careful slit lamp examination can also help identify signs of keratoconus including Munson sign, corneal scarring, Fleischer ring, Vogt striae and apical thinning. Early detection leads to better outcomes in these patients, especially if corneal crosslinking can slow the progression of the disease.[3]

Patients with disabilities deserve the same level of care as the rest of the population, and it is our ethical obligation as clinicians to provide such care. We should all be healthcare advocates for patients who cannot advocate for themselves.

Acknowledgement

Special thanks to Lindsay Sicks, OD, the attending doctor and mentor for this case.

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