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Unilateral implantation of a new non-diffractive extended range-of-vision IOL in a young patient with Curschmann-Steinert myotonic dystrophy

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ABSTRACT

Purpose: We present the case of a 36-year old Curschmann-Steinert myotonic dystrophy patient with posterior subcapsular cataract that we treated with unilateral implantation of an extended depth of focus intraocular lens to address his wish for spectacle independence at far and intermediate distance.

Observations: The patient underwent phacoemulsification with subsequent implantation of the AcrySof IQ Vivity IOL (Alcon, Fort Worth, TX, USA) in his left eye. Uncorrected distance visual acuity (UDVA) on the left eye increased from +0.40 logMAR preoperatively to −0.12 logMAR at 3 months postoperatively. At the three months follow-up distance corrected intermediate visual acuity (DCIVA) at 80 cm distance was −0.08 logMAR and DCIVA at 66 cm distance was 0.14 logMAR for the left eye. The defocus curve showed a functional defocus of 2.0 diopters at 0.2 logMAR or better, corresponding to the extended depth of focus. Dysphotopsia evaluation with a Halo & Glare simulator (Eyeland-Design Network GmbH, Vreden, Germany) revealed a very low level of photic phenomena.

Conclusions and Importance: Unilateral implantation of a new generation, non-diffractive extended depth of focus IOL was well tolerated and provided good functional results for far and intermediate distances. The patient reported a very low level of photic phenomena.

1. Introduction

Curschmann-Steinert myotonic dystrophy (MD1) is an autosomal dominant multisystemic condition which is caused by the expansion of an unstable cytosine guanine thymine (CTG) repeat in the protein kinase gene (DMPK).[1] The clinical manifestations of the condition and the age of onset depend on the size of the unstable CTG repeat. An increase of repeat size from generation to generation is possible and can lead to an earlier onset in children of MD patients.[1] The possible symptoms are diverse and include neuromuscular symptoms like muscular weakness and myotonia as well as cardiac conduction defects, respiratory insufficiency and endocrine disturbances.[2] Muscular atrophy can lead to a typical appearance referred to as 'facies myopathica' and a cachectic habitus. A receding hairline is another typical physical trait.[3] There are several known ocular manifestations: Most of the patients develop posterior subcapsular cataract that may be star-shaped at a young age. Incipient lens opacifications can be observed in many myotonic dystrophy type 1 (MD 1) patients in their third decade of life and lead to the development of visually significant cataract. Ptosis and lagophthalmos due to a weakness of the facial musculature can be present.[2,3] Maculopathy associated to MD1 has been reported[4] as well as capsulorhexis contraction after cataract surgery in patients suffering from MD 1[5]. The early onset of cataract can lead to the necessity to perform cataract surgery at a young age in those patients.

2. Case report

A 36-year old male patient presented to our clinic with bilateral posterior subcapsular cataract. The patient had been diagnosed with myotonic dystrophy type 1. The patient exhibited a number of typical symptoms of MD – myotonia and endocrine disturbances. Due to this condition he suffered from ptosis, lagophthalmus and dry eye syndrome. At the time of presentation, the patient was significantly disturbed by decreased visual acuity on the left eye. Uncorrected visual acuity on the right eye, however, was sufficient at all distances for him not to wear spectacles. Slit-lamp examination revealed a reduced tear break-up time in both eyes and bilateral posterior subcapsular cataract. Best corrected distance visual acuity (CDVA) was +0.30 logMAR for the right eye with

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a refraction of +0.25 diopters sphere (DS) −0.75 diopters cylinder (DC) x107° and +0.50 logMAR for the left eye with a refraction of +1.25 DS -0.75 DC x29°. Axial length was 23.36 mm for the right eye and 23.52 mm for the left eye measured with the IOL Master 700 (Carl Zeiss Meditec, Jena, Germany). Optical biometry revealed a corneal astigmatism of −0.93 D x 169° for the right eye and of −0.93 D x 11° for the left eye which was confirmed using the Pentacam HR tomography (Oculus GmbH, Wetzlar, Germany). Optical biometry was repeated to confirm these results three weeks after the first measurement and after treatment with artificial tears. The patient had a strong wish for spectacle independence especially for far and intermediate distance and was only willing to undergo surgery on the left eye.

The patient was informed in detail about different options of posterior chamber intraocular lenses (IOLs) including multifocal, Extended Depth of Focus (EDoF) and monofocal IOLs and about the possible benefits and complications related to the different options. The loss of accommodation and possible complications such as intraoperative posterior capsular rupture with the need to implant a monofocal IOL were discussed. Considering the patient's wishes for spectacle independence, the unilateral surgery and desire for only low amounts of glare and halos, we proposed the implantation of a new non-diffractive EDoF IOL to achieve spectacle independence at far and intermediate distance. After careful consideration, the patient decided on this option, as it matched best his requirements.

He underwent femtosecond laser-assisted phacoemulsification with a LenSx Laser (Alcon, Fort Worth, TX, USA) on the left eye and subsequent implantation of an AcrySof IQ Vivity IOL (Alcon, Fort Worth, TX, USA) with a calculated IOL power of +21.0 diopters.

The AcrySof IQ Vivity IOL is a single-piece hydrophobic acrylic IOL with an overall diameter of 13.0 mm and an optic zone diameter of 6.0 mm. The extension of the visual range to the intermediate region is achieved through non-diffractive Wavefront-Shaping Technology (X-Wave technology). The AcrySof IQ Vivity features the central 2.2-mm area with two transition elements. The first transition element stretches the wavefront resulting in continuous focal range, but the light is stretched in both directions, the myopic direction and the hyperopic direction; the light at hyperopic direction is behind the retina, therefore it is not very useful. Then the transition element two shifts the wavefront anteriorly, shifting the light from the hyperopic direction to the myopic direction, so that all the light energy is utilized. Recently, the manufacturer introduced a toric version of the IOL, but it was not available at the time of implantation.

Fig. 1 shows an intraoperative image. The main incision was placed at 101° using the Verion digital marking system (Alcon, Fort Worth, TX, USA) and an opposite clear-cornea incision was made at 281° after the IOL implantation to reduce the preexisting corneal astigmatism. The intra- and postoperative courses were uneventful.

At the three-months follow-up visit, UDVA of the left eye was −0.12 logMAR. The manifest refraction (MR) was 0.0 DS -0.25 DC x 35°. CDVA was −0.12 logMAR. Uncorrected intermediate visual acuity (UIVA) and distance corrected intermediate visual acuity (DCIVA) at 80 cm distance were both −0.08 logMAR. DCIVA at 66 cm distance was 0.14 logMAR. Uncorrected near visual acuity (UNVA) and distance corrected near visual acuity (DCNVA) at 40 cm were both 0.46 logMAR. Binocular UDVA was −0.10 logMAR, binocular UIVA (at 80 cm) was −0.08 logMAR and binocular UNVA (at 40 cm) was +0.10 logMAR. The defocus curve for the left eye is shown in Fig. 2. It shows a functional defocus of 2.0 diopters at 0.2 logMAR or better, illustrating the extended depth of focus from far to intermediate distance. Fig. 3 shows a slit-lamp photo of the postoperative result.

We evaluated photic phenomena using a Halo & Glare simulator (Eyeland-Design Network GmbH, Vreden, Germany). The simulator allows one to choose between three types of halo (classic halo, starburst and irregular halo). The patient was asked to adjust the size (from 0 to 100) and intensity (from 0 to 100) according to his own perception. For glare simulation, the simulator allows two different shapes (classic glare and asymmetric glare). The patient could adjust intensity and size of the glare parameter in the same way he could alter the halo. At the three-months examination, the patient reported a starburst type of halo, the size was 40 and the intensity 50. He did not report any form of glare. The result of the simulation is shown in Fig. 4.

The patient was very satisfied with the result and indicated that he would undergo the procedure again if he had to choose again. He reported full spectacle independence.

3. Discussion

We observed very good functional results in our patient for uncorrected intermediate and far visual acuity and a low level of photic phenomena.

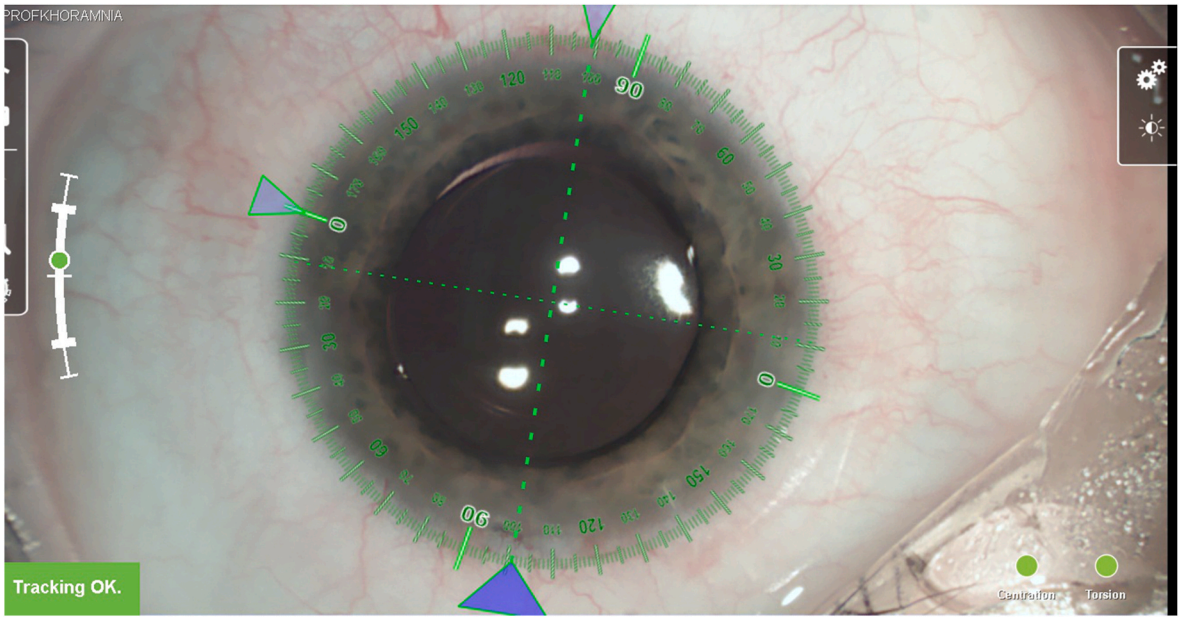

Fig. 1. Intraoperative image with displayed digital marking system. The main incision was placed at 101° (large arrow) and an opposite clear-cornea incision was placed 180° from the main incision at 281° (small arrow in the upper part of the image).

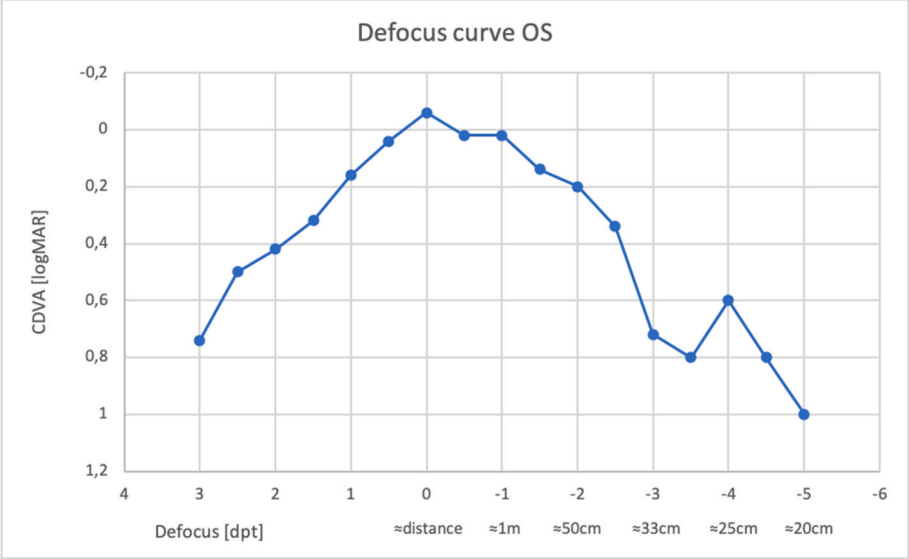

Fig. 2. Defocus curve of the left eye. The CDVA is 0.20 logMAR or better for a defocus of −2.0D to +1.0D.

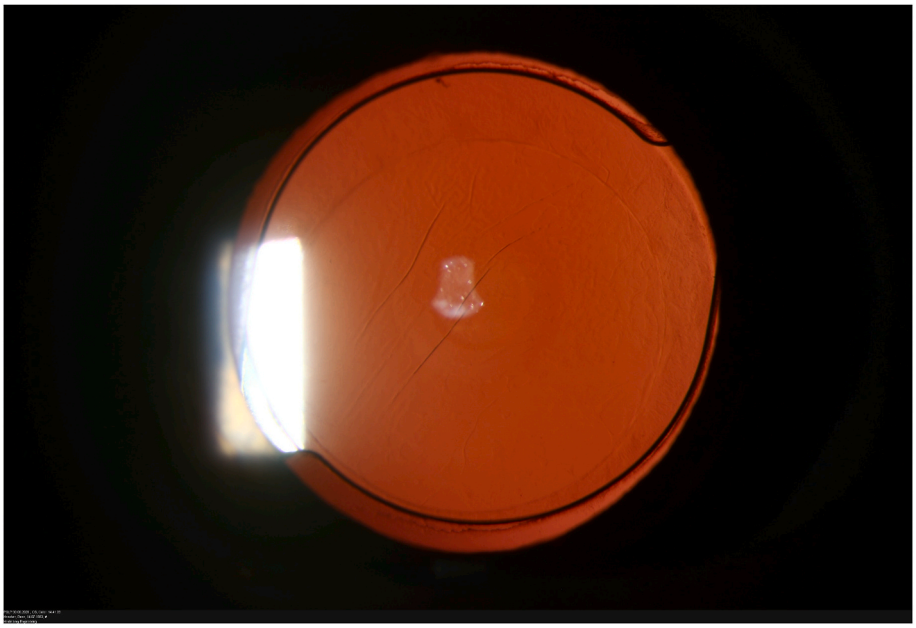

Fig. 3. Postoperative slit-lamp photograph of the AcrySof IQ Vivity IOL.

The patient was very satisfied with the outcome. However, when planning the surgery, we were faced with several challenges: The patient was only willing to undergo surgery on one eye and, as it is often the case in young patients, he was not used to wearing reading glasses or bifocals and therefore had a strong wish for spectacle independence. Furthermore, we had to anticipate potential complications associated with myotonic dystrophies, such as anterior capsule contraction.

Trifocal IOLs can provide good functional results for far, near and intermediate distances.[6,7,54] They are superior to monofocal IOLs at near and intermediate distance and provide better optical quality than bifocal IOLs at intermediate distance.[8,9] It has also been demonstrated that trifocal IOLs improve the near and intermediate reading acuity.[10] However, the data on unilateral trifocal IOL implantation are limited. Previous reports suggest, that optimal results can only be achieved after bilateral implantation of a multifocal IOL. Cionni et al. found significantly better results for near and intermediate distance after bilateral implantation of a diffractive multifocal IOL, the AcrySof SN60D3 ReSTOR IOL (Alcon, Fort Worth, TX, USA), compared to unilateral implantation of the same IOL. Patient satisfaction differed significantly between the two groups, with the patients who underwent bilateral implantation expressing higher overall satisfaction.[11] Häring et al. examined the differences in binocular functions after bilateral and unilateral implantation of a refractive multifocal lens and found significantly more distance and near aniseikonia after unilateral than after bilateral implantation.[12] Shoji et al. observed higher spectacle independence after bilateral implantation of a refractive multifocal IOL compared to unilateral implantation of the same IOL.[13] One of the limitations of diffractive technology is its spectral dependency, which may alter the IOL's performance under certain monochromatic light conditions.[14,15] Diffractive multifocal IOLs are also associated with high levels of dysphotopsia such as halo and glare and can reduce contrast sensitivity.[16] This arises from the optical principle of diffractive IOLs: in

Fig. 4. Result of the Halo & Glare simulation. The patient was asked to adjust the settings according to his own binocular visual impression, which consisted of the visual impression on the right eye with mildly pronounced cataract and the visual impression on the left eye with the AcrySof IQ Vivity IOL. The simulation revealed a very low level of photic phenomena.

which light is distributed between different foci and a superimposition of the images is created with only one image being in focus on the retina.[17] This explains the reduction of contrast sensitivity and the perception of halos. The scattering of the light from the diffractive rings can cause glare.[18] Interestingly, the study by Cionni et al. showed no statistically significant difference between the occurrence of halo and glare in patients after bilateral versus unilateral implantation of a diffractive multifocal IOL.[11] Thus, the unilateral implantation of a trifocal IOL has to be critically and cautiously assessed. When the patient compares the visual impressions of both eyes, he may not tolerate the photic phenomena or be dissatisfied with the visual acuity that is achieved after unilateral multifocal IOL implantation.

In this case, we chose to implant a non-diffractive EDoF IOL, in the expectation that there would be a better tolerance of the IOL being implanted in one eye only. EDoF IOLs extend the patient's visual range from far to intermediate distance. The extended depth of focus can be created by using different optical principles, including diffractive and refractive lens designs and small-aperture IOLs.[18–24] The AcrySof IQ Vivity IOL creates the extended focus range through X-Wave technology. As EDoF IOLs are a heterogenous category of IOLs, the American Academy of Ophthalmology has attempted to standardize an 'EDoF IOL' classification.[25] The criteria proposed include a CDVA that is non-inferior to a monofocal control IOL and a DCIVA that is superior to the control IOL as well as a depth of focus that is at least 0.5 D greater than for the monofocal control IOL.[25] A clinical study conducted for FDA approval confirmed these properties for the AcrySof IQ Vivity IOL, as well as similar contrast sensitivity and visual disturbances profile as the monofocal control IOL (AcrySof IQ IOL).[26] A meta-analysis by Liu et al. comparing trifocal (PanOptix [Alcon, Fort Worth, TX, USA], FineVison [PhysIOL, Liège, Belgium] and Lisa tri 839MP [Zeiss, Oberkochen, Germany]), EDoF (Tecnis Symfony ZXR00 [Johnson & Johnson, New Brunswick, New Jersey, USA]) and monofocal IOLs (Tecnis ZCB00 [Johnson & Johnson, New Brunswick, New Jersey, USA] and AcrySof SN60WF [Alcon, Fort Worth, TX, USA]) showed better results for the EDoF IOL (Tecnis Symfony ZXR00) at intermediate and near distance than for the monofocal IOLs, but the EDoF IOL (Tecnis Symfony ZXR00) was inferior to trifocal IOLs at near distance. The EDoF IOL (Tecnis Symfony ZXR00) showed reduced contrast sensitivity compared to the monofocal IOLs, but it showed better results for contrast sensitivity testing than trifocal IOLs. Symfony's spectral dependence was observed to affect visual acuity and contrast sensitivity.[14,27,28]

In contrast to other studies, this specific metaanalysis showed no difference between the EDoF IOL (Tecnis Symfony ZXR00) and the trifocal lenses regarding spectacle independence.[29] As EDoF IOLs feature different optical principles, the levels of dysphotopsia vary between the different IOLs. For certain EDoF IOLs such as the Mini Well (SIFI, Catania, Italy) a lower level of photic phenomena has been reported compared to trifocal IOLs.[30–32] Other studies found no differences in dysphotopsia between trifocal IOLs and the EDoF IOL Tecnis Symfony ZXR00.[29,33–35] According to the FDA safety and effectiveness data, the AcrySof IQ Vivity IOL is associated with a very low level of photic phenomena without differences compared to a monofocal aspheric IOL.[26]

It should be considered that the patient still had physiological accommodation in his fellow eye. This could, at least in part, be responsible for the good binocular UIVA and UNVA. We observed similar results in a young patient with traumatic cataract who underwent unilateral implantation of the same IOL.[36]

Due to his condition, our patient suffered from lagophtalmus and dry eye syndrome. It has been reported that patients suffering from dry eye syndrome have a higher tear osmolarity than the healthy population,[37] which leads to a significantly higher variability of keratometry measurements.[38] Inaccurate keratometry can lead to a high postoperative refractive error, which is the most common reason for dissatisfaction after multifocal IOL implantation.[39–41] Although it is known, that EDoF IOLs show a higher tolerance to residual refractive errors,[42] biometry was verified a few weeks after the first measurement to achieve the best possible outcome.

Several reports note a severe anterior capsular contraction in MD1 patients after cataract extraction.[5,43,44] A study by Ursell et al. found less capsular movement for the AcrySof MA60BM IOL (Alcon, Fort Worth, TX, USA) compared to a PMMA and hydrophilic IOL,[45] suggesting that the AcrySof IOL causes less capsular phimosis than these other IOLs. The AcrySof IQ Vivity IOL is based on the AcrySof IOL platform in that it is made of the same material and therefore a comparable risk for anterior capsular contraction for both IOLs might be assumed. Anterior capsular contraction can lead to IOL decentration.[46–49] The optical quality of bifocal and trifocal IOLs, however, depends strongly on a perfect centration of the IOL.[50–52] EDoF IOLs show a higher tolerance to decentration than multifocal IOLs,[53] which is of particular importance in a

case with an increased risk for anterior capsular contraction. So far, we did not observe capsular phimosis or IOL decentration in our patient.

4. Conclusion

The AcrySof IQ Vivity IOL provides good functional results for far and intermediate distance with a very low level of photic phenomena. Unilateral implantation was tolerated well.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Intellectual property

We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property.

Research ethics

We further confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

Declaration of competing interest

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