Ocular Axial Length in Unilateral Congenital Cataracts and Blepharoptosis
Gunter K. von Noorden and Richard Alan Lewis

Biometry performed in patients with unilateral dense congenital cataracts and unilateral complete blepharoptosis did not show a consistent elongation of the involved eye. The antero-posterior axis of the visually deprived eye was longer than that in the normal fellow eye in some but shorter or unchanged in other patients. These findings are in accord with the hypothesis gained from monkey experiments that visual deprivation during infancy may deregulate axial growth of the eye. However, unlike in monkeys where axial elongation usually prevails, this effect of visual deprivation is less predictable in humans.


Visual deprivation by lid suture, optical blurring, or chemical opacification of the cornea causes axial myopia in infant monkeys1-5 and cats6-9 as well as in lower animal species.10-12 However, the myopiogenic effect of lid closure has not always been consistent in monkeys13,14 and few comparable data about a possible relationship between visual deprivation and myopia exist from human studies. It has been reported, for instance, that eyes with corneal opacities become myopic,15 that aphakic refractive errors in congenital cataracts are shifted toward the myopic side,16 that a unilateral posterior polar cataract may be associated with elongation of the involved globe17 and that unilateral ptosis18,19 and even sleep19 cause axial myopia. On the other hand, refractive errors reported in other large series of patients with unilateral ptosis do not show any correlation between ptosis and myopia.20-22

In recognition of these controversies and the potential clinical importance of these mechanisms for the etiology of myopia, we measured the axial length of both eyes of patients with unilateral dense congenital cataracts unassociated with other detectable ocular anomalies and of both eyes of patients with unilateral complete congenital blepharoptosis.

Materials and Methods

Between 1978 and 1984, we studied ten patients who were older than six yr at examination, and had unilateral dense congenital cataracts in otherwise normal eyes. In all patients, the cataracts had been noted during the first year of life and reported to be complete, that is, obstructing the entire pupil. In eight patients, these cataracts were surgically removed between the ages of 2 and 4 yr (Cases 1–8) by irrigation and aspiration, using a single needle. In two patients, the unoperated cataracts were still in situ at the time of our examination and were totally opaque (Cases 9 and 10). None of the operated patients had implantation of a pseudophakos. Inclusion in this study required the fellow eye of each patient to be structurally normal with normal corrected visual acuity, and that there were no postoperative complications of the operated eye, such as secondary glaucoma. Each subject in the study underwent a complete ophthalmologic examination, and, to exclude microphthalmos, the horizontal and vertical corneal diameters were measured in all patients and found to be within normal limits with no asymmetry between paired eyes.

Two additional patients were examined who had photographically documented unilateral visual deprivation since birth from a complete ptosis associated with a congenital third nerve paralysis (Cases 11 and 12). In Case 11, the ptosis had been present for 53 yr; Case 12 had lid surgery at age 1½ yr, after having had complete ptosis since birth. Both patients were astigmatic and severely amblyopic in the involved eye.

Echographic biometry was performed on each eye of each subject with the Digital Biometric Ruler, Model DBR-300 (Sonometric Systems Inc., New York, NY). For its internal electronic calculations, the assumed velocity of sound in phakic globes was 1548 m/sec, and in aphakic globes 1532 m/sec. Measurements were performed on seated subjects, using the standard applanation device provided by the manufacturer. Five readings for the total axial length were obtained from each eye, and the average of each set of measurements was determined. The same examiner performed biometry on each eye of each subject, and was not informed of the objectives of this study. Standardized A-scan echographic biometry (Kretz Model 7200 MA, Kretztechnik, Austria) was also performed on all eyes. Because there was a high degree of agreement of these
measuring the axial variations from the axial length of the other eye, we obtained an estimate of this interocular variability between paired eyes by measuring the axial lengths of both phakic eyes in 686 consecutive patients over 45 years of age who were preparing to undergo cataract surgery in one eye. In this adult population, the examiner, method, and instrumentation were the same as in our primary study population. The mean axial length and standard error in this population for all right eyes (OD) was 23.65 mm ± 0.05 mm and 23.59 mm ± 0.07 mm for all left eyes (OS). In this same population, the mean difference of axial length (OS-OD) is 0.063 mm. Therefore if we wish to test the hypothesis that there is a difference between the axial length of a patient’s eyes at the 0.001 level, the absolute value of the axial length difference must be greater than 0.22 mm.

### Results

The data are summarized in Table 1. No significant difference existed between the axial lengths of the two eyes in Case 1. The axial length of the involved eye in patients with unilateral cataracts or a history of unilateral cataract were significantly longer than the normal eye in 7 out of 10 patients (Cases 2–6, 8, 9). The aphakic or cataractous eye was shorter than the normal eye in two other cases (Cases 7 and 10). Likewise, in both patients with complete congenital ptosis, the involved eye was shorter than the normal eye. Thus, our data do not support the notion of a consistent and predictable relationship between prolonged deprivation of patterned vision during infancy and axial elongation of the globe in humans.

### Discussion

Numerous observations support the conclusion that alterations of visual input in early life may affect axial growth of the eye in experimental animals and neural factors evoked by abnormal visual experience are thought to influence the growth of the posterior segment of the eye.

Patients with unilateral congenital cataracts appear to be ideal models to test this theory on the etiology of myopia in humans. The opaque lens deprives the eye presumably from birth of patterned vision by restricting retinal stimulation to that occurring from unpatterned diffuse light. Indeed, 7 of 10 eyes with this condition showed increased axial length of the involved eye. But the fact that no such effect occurred in one patient and that the involved eye was actually shorter than the control eye in two patients dilutes the significance of these findings.

It may be argued that Cases 7 and 10 represented variants of persistent hyperplastic primary vitreous (PHPV) which is, as a rule, associated with microphthalmos. In that case any axial elongation of the deprived eye would be obscured by the microphthalmos. However, the operative report of Case 7 contained no description of features commonly associated with PHPV and ultrasonic examination of the cataractous eye of Case 10 was negative for PHPV. Moreover, no difference existed between the corneal diameters in both eyes of our patients and the finding of a normal corneal diameter in the presence of microphthalmos is most uncommon.

It is also unlikely that cataract surgery may have influenced the axial length measurements. Not only can the findings in Case 10, in whom no surgery was performed, not be reconciled with this possibility, but there is no known observation or mechanism by which aspiration and irrigation of the lens through a single corneal needle puncture may cause elongation of the globe. It is of interest that Balacco-Gabrieli et al.

### Table 1. Biometry in unilateral cataract and ptosis

<table>
<thead>
<tr>
<th>Case</th>
<th>Condition</th>
<th>Eye</th>
<th>OD: Biometry</th>
<th>OS: Biometry</th>
<th>Diff.</th>
<th>Age at Exam</th>
<th>Age at Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Cataract</td>
<td>R</td>
<td>22.43 mm</td>
<td>22.62 mm</td>
<td>0.19 mm</td>
<td>7½ yr</td>
<td>4 yr</td>
</tr>
<tr>
<td>2</td>
<td>Cataract</td>
<td>R</td>
<td>22.80 mm</td>
<td>22.49 mm</td>
<td>0.31 mm</td>
<td>51 yr</td>
<td>2 yr</td>
</tr>
<tr>
<td>3</td>
<td>Cataract</td>
<td>R</td>
<td>22.25 mm</td>
<td>21.40 mm</td>
<td>0.85 mm</td>
<td>6 yr</td>
<td>6 mo</td>
</tr>
<tr>
<td>4</td>
<td>Cataract</td>
<td>R</td>
<td>22.40 mm</td>
<td>22.00 mm</td>
<td>0.40 mm</td>
<td>5½ yr</td>
<td>3 yr</td>
</tr>
<tr>
<td>5</td>
<td>Cataract</td>
<td>L</td>
<td>22.62 mm</td>
<td>24.55 mm</td>
<td>1.93 mm</td>
<td>15 yr</td>
<td>9 mo</td>
</tr>
<tr>
<td>6</td>
<td>Cataract</td>
<td>R</td>
<td>25.09 mm</td>
<td>23.86 mm</td>
<td>1.23 mm</td>
<td>13 yr</td>
<td>1½ yr</td>
</tr>
<tr>
<td>7</td>
<td>Cataract</td>
<td>L</td>
<td>21.76 mm</td>
<td>20.33 mm</td>
<td>1.53 mm</td>
<td>6 yr</td>
<td>1½ yr</td>
</tr>
<tr>
<td>8</td>
<td>Cataract</td>
<td>L</td>
<td>22.63 mm</td>
<td>24.00 mm</td>
<td>1.37 mm</td>
<td>15 yr</td>
<td>2 yr</td>
</tr>
<tr>
<td>9</td>
<td>Cataract</td>
<td>L</td>
<td>22.62 mm</td>
<td>26.20 mm</td>
<td>3.58 mm</td>
<td>25 yr</td>
<td>na</td>
</tr>
<tr>
<td>10</td>
<td>Cataract</td>
<td>L</td>
<td>24.70 mm</td>
<td>21.70 mm</td>
<td>3.00 mm</td>
<td>17 yr</td>
<td>na</td>
</tr>
<tr>
<td>11</td>
<td>Complete Ptosis</td>
<td>L</td>
<td>25.17 mm</td>
<td>24.08 mm</td>
<td>1.09 mm</td>
<td>53 yr</td>
<td>na</td>
</tr>
<tr>
<td>12</td>
<td>Complete Ptosis</td>
<td>R</td>
<td>22.50 mm</td>
<td>23.34 mm</td>
<td>0.84 mm</td>
<td>11 yr</td>
<td>1½ yr</td>
</tr>
</tbody>
</table>

### Visual Acuity

<table>
<thead>
<tr>
<th>OD: 20/20</th>
<th>OS: 20/200</th>
</tr>
</thead>
<tbody>
<tr>
<td>OD: 1/200</td>
<td>OS: 20/20</td>
</tr>
</tbody>
</table>

### Refraction

| -2.25 sph | -5.50 X 90 |
| +1.25 sph | +2.50 X 140 |
| +0.75 +0.50 X 90 |
served an aphakic refractive error of less than ten diopters and an increased anterior-posterior diameter of the eyes in 5 of 19 children with unilateral or bilateral congenital cataracts. However, since neither the raw data nor the basis of comparison are supplied by the authors, it is difficult to apply these findings to our study.

That elongation of the eye occurred in the normal rather than in the affected eye of two patients with complete congenital ptosis is not in accord with a consistent effect of visual deprivation on axial elongation. Moreover, this finding invites a critical review of clinical data that have been cited to establish an association between ptosis and myopia in humans.11 Unless a ptosis is complete, ie, obstructing the pupil so that the patient cannot see even by tilting the head backwards, the eye is not visually deprived. Hoyt et al10 reported myopia and increased axial length in 8 of 64 patients with complete unilateral congenital ptosis. Since the refractive errors or axial lengths in the remaining 56 patients are not mentioned, these findings are insufficient to support a causal relationship between lid closure and myopia. O’Leary and Milrodot11 found incomplete ptosis to be associated with myopia but three similar studies20–22 using larger patient samples failed to establish a consistent correlation between myopia and incomplete ptosis. Thus, no convincing evidence has been presented that complete or incomplete ptosis cause myopia in humans.

We conclude from our data that in patients with unilateral congenital cataracts or complete congenital unilateral blepharoptosis, biometry frequently shows differences in axial length between the involved and uninvolved eyes. These differences exceed the interocular difference that is present in a normal adult population. In patients with unilateral cataracts, the anteroposterior axis of the visually-deprived eye was longer than that in the fellow eye in most but was also shorter or unchanged in other patients.

Thus, the effect of visual deprivation on axial elongation of the eye seems less consistent in humans than in monkeys where most eyes respond to visual deprivation with axial elongation.5 In view of their significance for the etiology of refractive errors, these findings deserve further exploration by a multicenter collaborative study that is based on a larger patient sample. Such a study is being planned.

Key words: congenital cataract, congenital blepharoptosis, axial length of the eye, myopia, biometry, visual deprivation, refractive error

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References