The Timing for Diagnosis of Anisometropia in Japanese Children with Congenital Eyelid Ptosis

Toshihiko Matsuo1,2,*
1Department of Ophthalmology, Okayama University Hospital and Okayama University Medical School, Japan
2Okayama University Graduate School of Interdisciplinary Science and Engineering in Health Systems, Japan

Abstract

Purpose: The difference in refractive errors between both eyes, namely, anisometropia, is known complication in congenital blepharoptosis. This study aimed to elucidate the timing of diagnosis of anisometropia in children with congenital blepharoptosis.

Methods: Retrospective review was made on medical records of 20 consecutive patients with congenital blepharoptosis at Okayama University Hospital from 2004 to 2018.

Results: The age at the initial visit ranged from 1 month to 5 years 1 month (median, 5.5 months), and the follow-up period from 1 year 8 months to 13 years 5 months (median, 5 years 5 months). Blepharoptosis was on the right side in 6 patients, on the left side in 10, and on both sides in 4. Four patients underwent levator muscle plication at the age ranging from 3 months to 6 years 2 months: One patient on the right side, 2 on the left side, and one on both sides. Nine patients were found to have anisometropia, defined as hyperopic difference of 2 or more diopters between both eyes, or astigmatic difference of 2 or more diopters between both eyes, and astigmatism of 3 or more diopters in both eyes (anisometropia), at the age ranging from 1 year 3 months to 3 years 6 months (median, 3 years). Four of the 9 patients with anisometropia showed unilateral amblyopia as the best-corrected visual acuity in decimals ranging from 0.2 to 0.5. The remaining 11 patients without anisometropia had normal levels of visual acuity in both eyes. Anisometropia was found in 3 of 4 patients with surgery and 6 of 16 patients with no surgery (no significant difference at chi-square test).

Conclusion: About a half of children with congenital blepharoptosis was found to have anisometropia at the age of 1 to 3 years. Care must be taken on refraction and glasses prescription at these ages.

Keywords: Congenital blepharoptosis; Anisometropia; Levator muscle plication

Introduction

Congenital blepharoptosis, or eyelid ptosis, is usually an isolated condition of eyelid drooping involving unilateral side or both sides [1-4]. The hypoplastic levator muscle with uncertain genetic background is the underlying cause for congenital blepharoptosis, and complications with other systemic anomalies are rare. Other syndromic conditions such as congenital fibrosis of the extraocular muscles are not included in the entity of congenital blepharoptosis.

The eyelid drooping is present at birth and noted as blepharoptosis when an infant tries to open the eyes in a period of arousal. The extent of blepharoptosis varies from infant to infant, and eyelid-raising usually becomes better as the lapse of months after birth. The infants or children with blepharoptosis take chin-up posture to compensate the eyelid-drooping to make the pupillary area uncovered by the eyelid. When the pupillary area is covered consistently with drooped eyelid, the absence of visual input to the unilateral eye or both eyes will lead to deprivation amblyopia. Under the circumstances, surgical intervention to lift the eyelid has to be planned to avoid and reverse the state of amblyopia. In addition to deprivation amblyopia, the difference in refractive errors between both eyes, namely anisometropia, is a known complication in congenital blepharoptosis [5-10]. This study reviewed consecutive children with blepharoptosis, irrespective of surgical intervention, and aimed to elucidate the timing of diagnosis of anisometropia in children with congenital blepharoptosis.

Methods

Retrospective review was made on medical records of 24 consecutive patients with congenital blepharoptosis who were seen at Okayama University Hospital from 2004 to 2018. The study reviewed consecutive children with blepharoptosis, irrespective of surgical intervention, and aimed to elucidate the timing of diagnosis of anisometropia in children with congenital blepharoptosis.
Table 1: Summary of 20 consecutive children with congenital blepharoptosis.

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Gender/Age at initial visit</th>
<th>Follow-up period</th>
<th>Laterality</th>
<th>Surgery (Age)</th>
<th>Other anomalies</th>
<th>Best-corrected visual acuity (refractive error) in right eye at last visit</th>
<th>Best-corrected visual acuity (refractive error) in left eye at last visit</th>
<th>*Hyperopia/Astigmatism (age at onset)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Male/8 mo</td>
<td>1 yr 8 mo</td>
<td>Left</td>
<td>No</td>
<td>No</td>
<td>1.5 (-0.5D c-0.5DA90)</td>
<td>1.2 (+3.5D c-1.5DA40)</td>
<td>Yes (3 yr)</td>
</tr>
<tr>
<td>2</td>
<td>Male/2 mo</td>
<td>13 yr 5 mo</td>
<td>Left</td>
<td>No</td>
<td>No</td>
<td>1.5 (+0.5D c-0.5DA170)</td>
<td>1.0 (+4.5D c-1.5DA30)</td>
<td>Yes (3 yr 2 mo)</td>
</tr>
<tr>
<td>3</td>
<td>Male/1yr 5 mo</td>
<td>8 yr 4 mo</td>
<td>Left</td>
<td>Yes 1 yr 6 mo 3 yr 4 mo</td>
<td>No</td>
<td>1.5 (+1.5D)</td>
<td>1.0 (+2.25D c-2.0DA115)</td>
<td>Yes (3 yr 2 mo)</td>
</tr>
<tr>
<td>4</td>
<td>Male/5 yr 1 mo</td>
<td>3 yr 10 mo</td>
<td>Left</td>
<td>Yes 6 yr 2 mo</td>
<td>Craniosynostosis (No surgery)</td>
<td>1.2 (+1.25D c-1.0DA10)</td>
<td>1.5 (+0.5D)</td>
<td>No</td>
</tr>
<tr>
<td>5</td>
<td>Male/1 mo</td>
<td>11 yr 3 mo</td>
<td>Bilateral</td>
<td>Yes 3 mo 1 yr 9 mo</td>
<td>Lower eyelid entropion on left side</td>
<td>1.5 (-2.0D)</td>
<td>0.2 (-2.5D c-2.0DA25)</td>
<td>Yes (3 yr 3 mo)</td>
</tr>
<tr>
<td>6</td>
<td>Male/3 mo</td>
<td>6 yr 7 mo</td>
<td>Left</td>
<td>No</td>
<td>No</td>
<td>1.5 (-0.75D)</td>
<td>1.5 (nil)</td>
<td>No</td>
</tr>
<tr>
<td>7</td>
<td>Female/10 mo</td>
<td>7 yr 3 mo</td>
<td>Bilateral</td>
<td>No</td>
<td>No</td>
<td>1.5 (-0.75D)</td>
<td>1.5 (-0.75D)</td>
<td>No</td>
</tr>
<tr>
<td>8</td>
<td>Female/2 mo</td>
<td>8 yr 7 mo</td>
<td>Bilateral</td>
<td>No</td>
<td>VSD (spontaneous closure)</td>
<td>0.6 (+3.75D c-3.0DA170)</td>
<td>0.5 (+4.25D c-3.25DA180)</td>
<td>Yes (3 yr 6 mo)</td>
</tr>
<tr>
<td>9</td>
<td>Male/11 mo</td>
<td>4 yr 5 mo</td>
<td>Right</td>
<td>No</td>
<td>No</td>
<td>0.9 (+2.5D c-1.25DA90)</td>
<td>0.8 (+2.75D c-1.75DA90)</td>
<td>No</td>
</tr>
<tr>
<td>10</td>
<td>Male/1yr 1 mo</td>
<td>4 yr 10 mo</td>
<td>Right</td>
<td>No</td>
<td>Cleft lip and palate with 2 surgeries at 1 yr and 1 yr 6 mo</td>
<td>1.2 (-0.5D c-0.5DA20)</td>
<td>1.2 (-0.5D c-0.5DA180)</td>
<td>No</td>
</tr>
<tr>
<td>11</td>
<td>Male/1 yr 8 mo</td>
<td>1 yr 9 mo</td>
<td>Bilateral</td>
<td>No</td>
<td>No</td>
<td>0.6 (-0.5D)</td>
<td>0.6 (+0.5D)</td>
<td>No</td>
</tr>
<tr>
<td>12</td>
<td>Male/2 mo</td>
<td>6 yr</td>
<td>Right</td>
<td>Yes 8 mo</td>
<td>Lower eyelid entropion on right side</td>
<td>0.2 (+2.0D c-4.0DA80)</td>
<td>0.9 (+0.5D c-0.5DA50)</td>
<td>Yes (2 yr)</td>
</tr>
<tr>
<td>13</td>
<td>Female/7 mo</td>
<td>4 yr 10 mo</td>
<td>Right</td>
<td>No</td>
<td>Turner syndrome VSD (surgery at 1 yr 7 mo)</td>
<td>0.5 (+4.5D c-1.25DA130)</td>
<td>0.8 (+2.25D c-0.75DA130)</td>
<td>Yes (2 yr 4 mo)</td>
</tr>
<tr>
<td>14</td>
<td>Female/1 mo</td>
<td>6 yr 1 mo</td>
<td>Left</td>
<td>No</td>
<td>No</td>
<td>1.5 (+2.25D c-0.5DA180)</td>
<td>1.2 (+2.75D c-1.5DA160)</td>
<td>No</td>
</tr>
<tr>
<td>15</td>
<td>Male/5 mo</td>
<td>5 yr 7 mo</td>
<td>Right</td>
<td>No</td>
<td>No</td>
<td>1.0 (+2.0 c-1.0DA165)</td>
<td>1.0 (+1.5D c-0.75DA175)</td>
<td>No</td>
</tr>
<tr>
<td>16</td>
<td>Male/3 mo</td>
<td>5 yr 3 mo</td>
<td>Left</td>
<td>No</td>
<td>No</td>
<td>1.2 (+2.25 c-0.75DA135)</td>
<td>0.7 (+3.0 c-2.75DA180)</td>
<td>Yes (2 yr 1 mo)</td>
</tr>
<tr>
<td>17</td>
<td>Male/1 mo</td>
<td>1 yr 8 mo</td>
<td>Left</td>
<td>No</td>
<td>No</td>
<td>u.m. (+0.25D c-0.5DA175)</td>
<td>u.m. (+0.5D c-2.5DA175)</td>
<td>Yes (1yr 3 mo)</td>
</tr>
<tr>
<td>18</td>
<td>Female/1 yr</td>
<td>3 yr</td>
<td>Right</td>
<td>No</td>
<td>No</td>
<td>1.0 (+1.25D c-0.5DA5)</td>
<td>0.9 (+1.0 c-0.5DA45)</td>
<td>No</td>
</tr>
<tr>
<td>19</td>
<td>Female/1 yr 7 mo</td>
<td>2 yr</td>
<td>Left</td>
<td>No</td>
<td>No</td>
<td>1.0 (+1.0D c-0.5DA25)</td>
<td>1.0 (+1.0D c-0.5DA15)</td>
<td>No</td>
</tr>
<tr>
<td>20</td>
<td>Female/3 mo</td>
<td>2 yr 10 mo</td>
<td>Left</td>
<td>No</td>
<td>No</td>
<td>1.0 (-1.75D c-0.25DA115)</td>
<td>1.0 (-1.5D c-1.25DA25)</td>
<td>No</td>
</tr>
</tbody>
</table>

yr: year; mo: month; VSD: Ventricular Septal Defect; u.m.: unmeasurable

*Hyperopia/Astigmatism indicates 2 diopters or more hyperopic difference between both eyes, or 2 diopters or more astigmatic difference between both eyes or 3 diopters or more astigmatic errors in both eyes

conformed to the tenets of the declaration of Helsinki and was approved as a retrospective study by the ethics committee of Okayama University Graduate School of Medicine, Dentistry, and Pharmaceutical Sciences and Okayama University Hospital. Four of the 24 patients were excluded from the study; 2 patients with no follow-up, one patient with congenital facial nerve hypoplasia, and one patient with Down syndrome who could not cooperate with refractive error measurement at the age of 2 years 4 months at the last visit.

Results

The 20 consecutive patients (Table 1) were 13 boys and 7 girls. The age at the initial visit ranged from 1 month to 5 years 1 month (median, 5.5 months; mean, 9.9 months), and the follow-up period ranged from 1 year 8 months to 13 years 5 months (median, 5 years 5 months; mean, 71.5 months). The systemic complications were ventricular septal defect in 2 patients, cleft lip and palate in one, craniosynostosis in one. Blepharoptosis was noted on the right side in 6 patients, on the left side in 10, and on both sides in 4. Four patients underwent levator muscle plication at the age ranging from 3 months to 6 years 2 months (Table 1): one patient on the right side, 2 on the left side, and one on both sides. Two of 4 patients with surgery resulted in amblyopia: A boy (Case 5) underwent a first surgery for levator plication on both sides at the age of 3 months, and had a second surgery on both sides at the age of 1 year 9 months, ending up with the best-corrected visual acuity of 0.2 in the left eye, in contrast with 1.5 in the right eye. Another boy (Case 12) underwent levator plication on the right eye, ending up with the best-corrected visual acuity of 0.2. These two patients had also lower eyelid entropion in amblyopic eyes.

From a different perspective, 9 patients were found to have anisometropia, defined as hyperopic difference of 2 or more diopters between both eyes, or astigmatic difference of 2 or more diopters between both eyes, or astigmatism of 3 or more diopters in both eyes (ametropia), at the age ranging from 1 year 3 months to 3 years 6 months (median, 3 years; mean, 31.6 months). Four of the 9 patients with anisometropia showed unilateral amblyopia as the best-corrected visual acuity in decimals ranging from 0.2 to 0.5. Two patients with 0.2 and the other two with 0.5. The remaining 11 patients without anisometropia had normal levels of visual acuity in both eyes. Anisometropia was found in 3 of 4 patients with surgery and 6 of 16 patients with no surgery (no significant difference at chi-square test).

Discussion

The goal of this study is to reveal long-term outcome of visual
acuity in children with congenital blepharoptosis from the viewpoint of anisometric amblyopia in addition to deprivation amblyopia. This study is unique in the point that all children with diagnosis of congenital blepharoptosis, irrespective of surgical intervention, were included in the study population. The preceding studies on consecutive series of blepharoptosis usually included patients with surgeries [5-10] while other reports were population-based studies [1-3]. The standard indication for surgical intervention in congenital blepharoptosis is to open the pupillary area which is covered by the drooped eyelid, in order to avoid and reverse deprivation amblyopia. Anisometropia has been known to be present in congenital blepharoptosis and been considered to be caused by abnormal movement of upper eyelids. The corneal shape would be influenced by the movement of upper eyelids and thus, astigmatism would be induced in blepharoptosis [11]. The axis of astigmatism might be changed after surgical intervention [12-18]. In general, anisometropia in itself should not be made as the reason for surgical intervention in congenital blepharoptosis [19].

In this study population, two children with surgery (Case 5 and 12) showed as dense anisometropia as the best-corrected visual acuity of 0.2 in one eye. These two patients had, in addition, lower eyelid entropion in amblyopic eyes, and care for superficial punctate keratopathy was required in the follow-up. The corneal problem might also contribute to dense anisometropia in these two patients. It should be noted that levator plication was combined with lower eyelid entropion surgery as a second surgery in the other eye of Case 5 with bilateral blepharoptosis, resulting in good visual acuity. Refractive errors can be measured certainly in children at the age of 1 year or older with a hand-held refractometer or table-fixed refractometer. In the standard treatment for anisometropia or ametropia, glasses with full correction were prescribed according to cycloplegic refractive errors which were determined by topical application of 1% cyclopentolate. All patients in this study population gained the best-corrected visual acuity of 0.5 or better except for two children with the visual acuity of 0.2 in one eye. A major limitation in this study is the small number of children with congenital blepharoptosis at a single institution.

In the field of pediatric ophthalmology, congenital blepharoptosis is relatively frequent among congenital diseases such as congenital cataract [20], ocular surface dermoid [21], ectopia lentis (lens subluxation) [22], and congenital superior oblique muscle palsy [23]. Clinical decision upon the observation or surgical intervention and the timing for surgical intervention is a key for following these children with congenital diseases to have them obtain better visual acuity. In the follow-up, checking and correction of refractive errors is mandatory in children who are in the critical period of visual development, as shown in the present study.

**Conclusion**

About a half of children with congenital blepharoptosis, irrespective of surgical intervention, was found to have anisometropia at the age of 1 to 3 years in this study population. Care must be taken on refraction and glasses prescription at these ages.

**References**