Anisometropia and Ptosis in Patients with Monocular Elevation Deficiency

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ABSTRACT
Objective: To determine the effect of ptosis on the refractive error in eyes having monocular elevation deficiency.
Study Design: Case series.
Place and Duration of Study: Al-Shifa Trust Eye Hospital, Rawalpindi, from January 2011 to January 2014.
Methodology: Visual acuity, refraction, orthoptic assessment and ptosis evaluation of all patients having monocular elevation deficiency (MED) were recorded. Shapiro-Wilk test was used for tests of normality. Median and interquartile range (IQR) was calculated for the data. Non-parametric variables were compared, using the Wilcoxon signed ranks test. P-values of <0.05 were considered significant.
Results: A total of 41 MED patients were assessed during the study period. Best corrected visual acuity (BCVA) and refractive error was compared between the eyes having MED and the unaffected eyes of the same patient. The refractive status of patients having ptosis with MED were also compared with those having MED without ptosis. Astigmatic correction and vision had significant difference between both the eyes of the patients. Vision was significantly different between the two eyes of patients in both the groups having either presence or absence of ptosis (p=0.04 and p < 0.001, respectively).
Conclusion: Significant difference in vision and anisoastigmatism was noted between the two eyes of patients with MED in this study. The presence or absence of ptosis affected the vision but did not have a significant effect on the spherical equivalent (SE) and astigmatic correction between both the eyes.


INTRODUCTION
Congenital incomitant types of strabismus, in which the magnitude varies with the position of gaze, are less common than the comitant types, where the size of deviation remains the same with any direction of gaze.1 It has been proposed to include monocular elevation deficiency (MED), an incomitant strabismus, under congenital innervation dysgenesis syndrome.2 In MED, there is limitation of elevation of the eye in both adduction and abduction. It may be associated with ipsilateral ptosis or pseudoptosis with or without a jaw winking phenomenon.3 Surgical treatment depends on the amount of hypotropia and horizontal deviation. Ptosis, if present, is treated later. Refractive error and amblyopia treatment is prescribed as is done while treating any type of squint. Prevalence of amblyopia or strabismus has been reported in different populations.4-6 Refractive errors in specific conditions have been investigated.7 Whether the main vertical pull on the globe in cases of tight inferior rectus in MED has any affect on the refractive error, needs attention of the ophthalmologist. Similarly, it is of interest to evaluate any possible effect of ptosis on the refractive error; and consequently, the vision when compared to the unaffected contra lateral eye.

The objective of this study was to compare the best corrected visual acuity (BCVA) and refractive error between the affected and unaffected eyes of patients with MED; and evaluate the effect of presence or absence of ptosis on refractive error and vision.

METHODOLOGY
This study included all patients of MED, observed from January 2011 to January 2014 at Al-Shifa Trust Eye Hospital, Rawalpindi. The study was conducted after approval from the Ethical Review Board of the institution. Informed consent was taken from the patients according to a protocol conforming to the Declaration of Helsinki. This research was not funded by any agency.

Consecutive sampling was done during the time period of the study. Inclusion criteria were all consecutive patients of MED presenting at a tertiary care eye centre. MED was described as limitation of elevation of the affected eye in both adduction and abduction. Patients having any organic cause of decreased vision on anterior and posterior segment examination of one or both eyes were excluded from the study. Patients with a history of ocular surgery were also excluded.

Detailed orthoptic assessment and ocular examination of the anterior and posterior segment was recorded.
Refractive error and BCVA were noted. Visual acuity was tested by means proper for age and then recorded as Snellen or Snellen-equivalent. The best corrected vision was recorded before the start of any amblyopia therapy. Comparison of BCVA between two eyes of the patients was done by converting to the logarithm of the minimum angle of resolution (logMAR). Refractive status was determined by cycloplegic refraction using 1% cyclopentolate eye drops. Spherical equivalent (SE) was recorded in case of astigmatism. Astigmatic refraction was also recorded. Amblyopia was recorded as BCVA below logMAR 0.7 (6/30). The findings were compared between the two eyes of each patient. Detailed ptosis examination was also recorded. Statistical analysis was performed using Statistical Package for Social Sciences (SPSS, Inc., Chicago, IL). Shapiro-Wilk test was used for tests of normality. Median and interquartile range (IQR) was calculated for the data. Non-parametric variables were compared using the Wilcoxon signed ranks test. P-values of < 0.05 were considered significant.

RESULTS

Forty-one patients, having monocular elevation deficiency, were included in the study. Twenty-three (56.1%) were females and 18 (43.9%) were males. Age of the patients ranged from 3 to 32 years (median 8.0, IQR 8). Spherical equivalent (SE) refractive error in eyes with MED ranged from -8.0 D to +7.0 D (median 0.50, IQR 2.25) while in the uninvolved fellow eye this range of SE was -6.0 D to +7.0 D (median 0.50, IQR 1.75). The difference in SE between the two eyes ranged from -5.0 to +5.5 (median 0.00, IQR 2.00). Astigmatism in eyes with MED ranged from -5.0 to 0.0 DC (median=0.00, IQR 1.00). The fellow eye had astigmatism between -3.00 to 0.0 DC (median 0.00, IQR 0.00). Difference in astigmatism between the involved and unaffected eye ranged from -4.00 to 1.50 DC (median 0.00, IQR 0.25). LogMAR vision in MED affected eyes ranged from -0.3 to 1.0 (median=0.00, IQR 0.25) and in the fellow eyes, the vision was recorded between -0.0 to 1.0 (median 0.20, IQR 0.35). The difference in vision between the two eyes ranged from -0.5 to 1.0 (median 0.00, IQR 0.60). Four patients had better vision in the affected eye compared with the uninvolved eye. Twelve (29.3%) patients had equal vision in both the eyes. The SE was not significantly different but astigmatic refraction and vision had significant difference among both the eyes (p=0.39, p=0.03 and p<0.001, respectively). The presence or absence of ptosis was also documented in these cases and the SE, astigmatic refraction, and vision was analysed in eyes with ptosis compared with those without ptosis. Twenty-eight (68.3%) patients had MED with ptosis. Jaw winking phenomenon was noticed in 18 (43.9%) patients.

Amblyopia recorded as BCVA below logMAR 0.7 (6/30) in the worst affected eye was seen in 14 (34.14%) patients. Among these 14 patients having amblyopia, 10 (71.42%) patients had ptosis associated with MED while 4 (28.57%) did not have ptosis.

SE was not significantly different between both eyes of the patients with and without ptosis (p=0.42 and p=0.82, respectively). Astigmatic refraction was not significantly different in patients without ptosis (p=0.59) but those having associated ptosis had significant difference in astigmatic refractive error (p=0.03) compared to fellow eyes. Vision, however, was significantly different in both the groups whether having ptosis or not (p=0.001 and p=0.04, respectively).

In patients having ptosis, 16 (57.1%) had MED in the right eye and 12 (42.9%) had MED in the left eye. In patients without ptosis, 4 (30.8%) had MED in the right eye and 9 (69.2%) had MED in the left. Most patients had hypermetropia or low myopic refractive error. Thirty-four patients (82.92%) with MED had either hypermetropic or less than -1.00D of myopic SE (Table I).

DISCUSSION

There is a likely association of MED with ptosis or pseudoptosis, with or without jaw winking phenomenon. Since quite a number of these patients are seen in our setup, the present study investigated the refractive error as well as visual status and compared it with the two eyes of patients having MED. Effect of ptosis, a known amblyogenic risk factor, was also evaluated.

Table I: Features of patients with monocular elevation deficiency.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Eyes with MED</th>
<th>Unaffected fellow eyes</th>
<th>MED with ptosis</th>
<th>MED without ptosis</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N (%)</td>
<td></td>
<td>N (%)</td>
<td>N (%)</td>
</tr>
<tr>
<td>SE</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Hypermetropia ≥ 1DS</td>
<td>17 (41.5)</td>
<td>17 (41.5)</td>
<td>9 (32)</td>
<td>5 (38.5)</td>
</tr>
<tr>
<td>&lt; 1 to &lt;=1DS</td>
<td>17 (41.5)</td>
<td>18 (44)</td>
<td>14 (50)</td>
<td>6 (46)</td>
</tr>
<tr>
<td>Myopia ≥1DS</td>
<td>7 (17)</td>
<td>6 (15)</td>
<td>5 (18)</td>
<td>2 (15.5)</td>
</tr>
<tr>
<td>Astigmatism</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt; 0.5 DC</td>
<td>26 (63)</td>
<td>32 (78)</td>
<td>16 (57)</td>
<td>10 (77)</td>
</tr>
<tr>
<td>0.5DC or &gt;</td>
<td>15 (37)</td>
<td>9 (22)</td>
<td>12 (43)</td>
<td>3 (23)</td>
</tr>
<tr>
<td>LogMAR vision</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt; 0.7</td>
<td>12 (29)</td>
<td>2 (5)</td>
<td>8 (28.5)</td>
<td>4 (31)</td>
</tr>
<tr>
<td>0-0.7</td>
<td>29 (71)</td>
<td>39 (96)</td>
<td>20 (71.5)</td>
<td>9 (69)</td>
</tr>
</tbody>
</table>

SE = Spherical equivalent, MED = Monocular elevation deficiency.
Studies have quantified and reported the increased risk of amblyopia with increasing SE and cylindrical anisometropia\(^8,9\). Strabismus related association of amblyopia has been reported in a recent study.\(^10\) The authors suggested larger studies to be carried out in patients with different types of strabismus to find the effects of extraocular muscle tension or lack thereof on the globe. This can help in detecting changes in the refractive error, so that these changes are timely addressed, if needed.\(^10\)

Reports are also available on the results of surgical management of many patients of MED.\(^11\) Some patients with ptosis tend to have astigmatic refractive error.\(^12\) Patients with congenital ptosis have higher rate of amblyopia due to greater prevalence of strabismus and refractive errors, although stimulus deprivation amblyopia was less commonly found in a study by Thapa.\(^12\) Another study stated that patients with congenital ptosis have the risk of anisometropic and strabismic amblyopia.\(^13\) Srinagesh and co-authors reported presence of amblyopia in 23.9% of eyes with more severe ptosis in cases with coexisting anisometropia or strabismus.\(^13\) A retrospective study over a 40-year period did not find any isolated vertical deviation in the patients having strabismus with childhood ptosis.\(^14\) There was no case of MED among the patients of childhood ptosis having strabismus in the above study.\(^14\)

Another study reported that unilateral simple congenital ptosis appears to have a predominance of left eye involvement.\(^15\) Almost equal frequency of laterality of ptosis with MED was seen in a report of 22 patients.\(^3\) The right eye was slightly more frequently involved in MED with ptosis (n=16), whereas 12 left eyes with MED had ptosis in this study.

Vision in children having MED ranged from logMAR 1.77 (20/1200) to logMAR 0.17 (20/30) and that in adults ranged from logMAR 1.47 to 0.17 (20/600 to 20/30) in a study of 28 patients.\(^16\) In comparison, better visual acuity range of 0.00 (6/6) to 1.0 (6/60) was recorded in this study. The vision was recorded after prescription of glasses where needed before the start of any amblyopia treatment. There may, however, be a potential for improvement in vision after amblyopia therapy.

There were four patients in which the eye having monocular elevation deficiency had better vision than the fellow eye in this study. This observation may be the consequence of fixation preference for the affected eye in certain cases of MED, similar to that reported in Duane syndrome.\(^17\) In each of these patients, there was a difference of 0.1, 0.2, 0.3 and 0.5 logMAR between the BCVA of the two eyes. A mixed etiology of refractive and strabismic amblyopia may occur. Stimulus deprivation due to severe ptosis has been reported in 12% of patients in a study after excluding the refractive and strabismic factors.\(^18\) Another study reported amblyopia in 14.9% of patients, mostly having unilateral childhood ptosis except for one patient having bilateral ptosis due to myasthenia gravis.\(^19\) The present study shows that presence of ptosis does not significantly affect the SE or astigmatic refraction, although vision is significantly different in the two eyes of MED patients. Higher frequency of amblyopia, however, was noted in MED patients having ptosis than those without (10 vs. 4).

A study reported amblyogenic ametropia in 32.1% of patients who received treatment for unilateral ptosis. Most of these had astigmatism greater than 2.50 diopters of cylinder.\(^20\) It is observed that children having ptosis with amblyopia due to anisoaustigmatism or deprivation may benefit from early surgery.\(^21\) This study showed a greater frequency of hypermetropic SE, more so in the MED patients having no ptosis (Table I). The authors have not found any report in the literature addressing the refractive error in MED compared to presence or absence of ptosis. The visual acuity needs to be screened early for any possible amblyopia therapy along with surgical correction of strabismus. Pediatric ophthalmologists need to detect refractive error and visual loss in the non-preferred eye to start early amblyopia therapy, as is preferably done in all cases of strabismus.

**CONCLUSION**

Significant difference in vision and anisoaustigmatism was noted between the two eyes of patients with MED in this study. Amblyopia was recorded more in MED patients having ptosis than in those without. Presence of ptosis, however, did not have a significant effect on the spherical equivalent (SE) and astigmatic correction between both the eyes.

**REFERENCES**


