Effect of Horizontal Rectus Surgery on Clinical and Paraclinical Indices in Congenital Nystagmus

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Purpose: To determine the effect of horizontal rectus muscle surgery on visual acuity, head posture and electronystagmographic indices in patients with congenital nystagmus.

Methods: This prospective comparative case series was conducted on 58 patients with congenital nystagmus over a period of three years. Patients were divided into three groups: the first group (29 cases) had head posture less than 20°, binocular visual acuity (BOVA) less than 20/30 and tropia less than 30Δ and underwent large recession of all four horizontal rectus muscles; the second group (23 cases) had head posture less than 20°, BOVA< 20/30 and tropia more than 30Δ who underwent large recession of two horizontal rectus muscles; and the third group (6 cases) had head posture more than 20° with any BOVA or tropia who underwent Kestenbaum-Anderson surgery.

Results: Mean age of the patients was 18.7±9.1 years and mean follow-up period was 17.5±7.4 months. Visual acuity improved in all three study groups and was statistically significant in the 2-rectus group (P<0.001). The speed and amplitude of nystagmus waves decreased in all groups which was statistically significant in the 4-rectus group (P values, 0.02 and 0.04, respectively). A small myopic shift was seen in the 2-rectus and 4-rectus groups and a small hyperopic shift was found in the Kestenbaum-Anderson group. Statistically significant improvement was achieved in eye deviation in the 2-rectus group and in head posture in the Kestenbaum-Anderson group (P<0.001).

Conclusion: Horizontal recti surgery in congenital nystagmus can improve visual acuity, ocular deviation and abnormal head posture, which is particularly marked with 2-rectus recession. Electronystagmographic indices improve especially with 4-rectus recession.


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INTRODUCTION

Nystagmus is an involuntary rhythmic conjugate oscillatory movement of the eyeballs with a slow component and is classified as jerky or pendular based on the velocity of movements, and as sensory or motor and congenital or acquired based on etiology. Visual loss in patients with nystagmus is attributed to short foveation time. Coexistence of macular or optic nerve abnormalities may aggravate the visual impairment in these eyes. In some patients, the intensity of the movements is decreased in certain positions of the head and the patient takes an abnormal head posture to obtain the best possible visual acuity.
Reduction of nystagmus frequency or amplitude by means of medical or surgical methods can theoretically increase foveation time, correct the abnormal head posture and improve cosmesis. After surgical correction of nystagmus, patients may experience subjective visual improvement despite unchanged visual acuity. Numerous medical and surgical treatments have been applied for nystagmus with abnormal head posture but there is less agreement on the treatment of nystagmus without a null point. Based on the results of the above-mentioned studies and our previous study, we planned a prospective comparative case series for treatment of different types of congenital nystagmus.

METHODS

This study was conducted on patients with congenital nystagmus who were referred to Labbafinejad Medical Center from 2002 to 2005. Subjects with previous extraocular muscle surgery and those less than 4 years of age were excluded. All individuals underwent a comprehensive examination including measurement of stereoacuity using the Titmus test, contrast sensitivity, worth-4-dot (W4D) test for far and near, monocular and binocular visual acuity for far and near, cycloplegic refraction, assessment of ductions and versions, evaluation of heterotropia using alternate cover test, head posture, anterior and posterior segment examination and electronystagmography. The ETDRS chart was used at 6 m for far visual acuity and the Snellen chart was used at 33 cm for near visual acuity measurement. Best-corrected monocular visual acuity was determined using positive lenses (3 diopters higher than the refractive error). Head posture was evaluated using a deviometer or torticulometer while the patient focused on an accommodative target at 6 m and 33 cm and was recorded as the degree of deviation from midline. All pre- and postoperative examinations were performed by an experienced ophthalmologist other than the surgeon under the same setting and lighting conditions.

In patients younger than 10 years, any refractive error was corrected and a 3-6 months period of amblyotherapy was carried out preoperatively to eliminate any interference from possible postoperative visual improvement due to correction of ametropia or amblyopia. For this purpose, all patients in this age group underwent cycloplegic refraction following twice instillation of cyclopentolate 1% and phenylephrine 5% drops, within a 5 minute interval. Subjective refraction was performed one week after cycloplegic refraction.

Based on the presence or absence of afferent pathway disorders such as albinism and macular or optic nerve disorders, patients were categorized into subgroups of sensory or motor nystagmus. The patients were divided into 3 groups considering the degree of head posture, binocular visual acuity (BOVA) and ocular deviation (tropia) as follows:

1) Patients with abnormal head posture <20°, BOVA <20/30 and tropia <30° underwent large recession of all four horizontal rectus muscles (4-rectus group).
2) Patients with abnormal head posture <20°, BOVA <20/30 and tropia >30° underwent large recession of two horizontal rectus muscles (2-rectus group).
3) Patients with abnormal head posture >20° with any BOVA or tropia underwent the Kes-tenbaum-Anderson procedure (KA group).

All operations were performed by one surgeon (AB). Patients were reexamined on days 1 and 7; months 1, 3, and 6, and every six months thereafter. Follow-up examinations included monocular and binocular visual acuity for far and near, Titmus test, W4D test for far and near, evaluation of head posture and electronystagmography. Subjective outcomes of surgery were also evaluated. In the KA group the results of surgery were also classified as excellent (abnormal head posture ≤5°), fair (abnormal head posture 5 to 15°) and poor (abnormal head posture >15°) based on final head posture. Abnormal head posture more than 5° in the opposite direction was considered as over-correction. Mean values were compared using t-test and frequency values were com-
pared using Chi-square or Fisher’s exact tests with significance set at P<0.05.

RESULTS

Overall 58 patients including 35 male (60.3%) and 23 female (39.7%) subjects with mean age of 18.7±9.1 (range 5-48) years were operated and followed for a mean period of 17.5±7.4 (range 6-36) months. Twenty-five patients (43.1%) had sensory disorders including oculo-cutaneous albinism in 22 cases (88%) and aniridia, optic atrophy and high myopia one case each (4%). Thirty-three patients (56.9%) had congenital motor nystagmus. Surgical interventions included large recession of all four horizontal rectus muscles in 29 cases (4-rectus group), large recession of two horizontal rectus muscles in 23 cases (2-rectus group) and the Kestenbaum-Anderson procedure in 6 cases (KA group).

Table 1 summarizes visual outcomes of surgery based on sensory or motor basis of the nystagmus. Far monocular vision (FMOV) was significantly improved in both sensory and motor groups but improvement in far binocular vision (FBOV) was significant only in the sensory group. Mean near monocular vision (NMOV) showed a decrease in both groups. Overall, changes in visual acuity included 0.03±0.2 logMAR in FBOV (P=0.3), 0.05±0.2 logMAR in near binocular vision (NBOV) (P=0.9), 0.1±0.2 logMAR in FMOV (P<0.001) and -0.04±0.3 logMAR in NMOV (P=0.1).

Contrast sensitivity was evaluated in 31 patients which revealed no change in 23 cases (74.2%) of whom 18 subjects (78.3%) subjectively reported improved visual quality. Contrast sensitivity improved in 8 (25.8%) cases, which was accompanied by subjective improvement in visual quality in 4 (50%) cases. No significant correlation was observed between improvement in contrast sensitivity and subjective improvement in visual quality (Fisher’s exact test, P=0.2).

Titmus test was applicable in 56 patients; 48 patients (85.7%) had no stereoaucity preoperatively, of whom 9 subjects (18.75%) gained some degree of stereoaucity. Patients who had stereoaucity preoperatively achieved significant improvement (McNemar test, P=0.004).

W4D test was applicable in 56 patients; 21 cases (37.5%) had fusion preoperatively, however 35 cases (62.5%) suffered from lack of fusion of whom 9 subjects (25.7%) gained fusion postoperatively (McNemar test, P= 0.004).

Changes in visual acuity, refractive errors and electronystagmographic indices based on surgical groups are shown in tables 2 and 3. Changes in abnormal head posture and tropia, as well as changes in frequency, amplitude and speed of nystagmus are presented in figures 1 to 5.

Table 1 Mean improvement in visual acuity based on sensory or motor basis of nystagmus

<table>
<thead>
<tr>
<th></th>
<th>Mean ± standard deviation (logMAR)</th>
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<tbody>
<tr>
<td></td>
<td>Sensory</td>
<td>Motor</td>
<td>Sensory</td>
<td>Motor</td>
</tr>
<tr>
<td>FBOV</td>
<td>0.06±0.1</td>
<td>0.006±0.25</td>
<td>0.005</td>
<td>0.9</td>
</tr>
<tr>
<td>NBOV</td>
<td>0.04±0.2</td>
<td>0.006±0.25</td>
<td>0.005</td>
<td>0.3</td>
</tr>
<tr>
<td>FMOV</td>
<td>0.11±0.22</td>
<td>0.001</td>
<td>0.02±0.2</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>NMOV</td>
<td>-0.04±0.23</td>
<td>0.3</td>
<td>-0.05±0.33</td>
<td>0.2</td>
</tr>
</tbody>
</table>

FBOV, far binocular vision; NBOV, near binocular vision; FMOV, far monocular vision; NMOV, near monocular vision

* t-test

K.A., Kestenbaum-Anderson

Figure 1 Mean values of head posture in the three study groups
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Figure 2 Mean tropia in the three study groups

Figure 3 Mean frequency of nystagmus in the three study groups

Figure 4 Mean values of nystagmus amplitude in the three study groups

Figure 5 Mean values of nystagmus speed in the three study groups
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Table 2 Mean and standard deviation for visual acuity (logMAR) and refractive errors (diopters) in the three study groups

<table>
<thead>
<tr>
<th></th>
<th>4-rectus group</th>
<th>2-rectus group</th>
<th>Kestenbaum-Anderson group</th>
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<tbody>
<tr>
<td></td>
<td>Pre-op</td>
<td>Post-op</td>
<td>P value*</td>
</tr>
<tr>
<td>FBOV</td>
<td>0.63±0.33</td>
<td>0.65±0.32</td>
<td>0.7</td>
</tr>
<tr>
<td>NBOV</td>
<td>0.66±0.44</td>
<td>0.63±0.40</td>
<td>0.4</td>
</tr>
<tr>
<td>FMOV</td>
<td>0.78±0.35</td>
<td>0.66±0.33</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>NMOV</td>
<td>0.54±0.35</td>
<td>0.63±0.36</td>
<td>0.04</td>
</tr>
<tr>
<td>Astigmatism</td>
<td>2.07±1.36</td>
<td>-2.24±1.47</td>
<td>0.1</td>
</tr>
<tr>
<td>SE</td>
<td>-0.75±0.25</td>
<td>-0.91±4.09</td>
<td>0.2</td>
</tr>
</tbody>
</table>

FBOV, far binocular vision; NBOV, near binocular vision; FMOV, far monocular vision, NMOV, near monocular vision; SE, spherical equivalent

*Paired t-test

Table 3 Mean and standard deviation for changes in electronystagmographic indices in the three study groups

<table>
<thead>
<tr>
<th>Changes in:</th>
<th>4-rectus P value*</th>
<th>2-rectus P value*</th>
<th>Kestenbaum-Anderson P value*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frequency (cycle/sec)</td>
<td>-0.47±1.7</td>
<td>0.2</td>
<td>0.25±0.88</td>
</tr>
<tr>
<td>Amplitude (degree)</td>
<td>-0.76±1.93</td>
<td>0.04</td>
<td>0.13±2.3</td>
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<tr>
<td>Speed (degree/sec)</td>
<td>-8.3±18.6</td>
<td>0.02</td>
<td>0.48±14.33</td>
</tr>
</tbody>
</table>

* Paired t-test, negative mark indicates decrease

The 4-Rectus Group

This group included 17 male (58.6%) and 12 female (41.4%) subjects with mean age of 19.4±6.9 (range 7-33) years. Nystagmus was motor in 17 (58.6%) and sensory in 12 (41.4%) cases. Mean duration of follow-up was 16.8±6.9 (range 6-31) months. Mean amount of muscle recession was 12.1±1.0 for bimedial recession (BMR) and 15.2±0.9 mm for bilateral recession (BLR) in this subgroup.

Changes in FBOV and NBOV were not statistically significant in this group, whereas mean FMOV increased and mean NMOV decreased significantly after the operation (table 2). Quality of vision improved in 23 (79.3%) and remained unchanged in 6 (20.7%) cases, however there was no significant correlation between subjective improvement in quality of visual and increased binocular or monocular visual acuity.

Mean spherical equivalent (SE) showed a mild but non-significant myopic shift postoperatively (table 2). Mean astigmatism also changed by a mean of -0.16±0.90 D (P=0.1) postoperatively which was with-the-rule (WTR) in 30 eyes, against-the-rule in 14 eyes and unchanged in 14 eyes (P=0.008).

Frequency, amplitude and speed of nystagmus all decreased in this group (table 3). Changes in electronystagmographic indices did not correlate with improvement in FBOV, NBOV or FMOV; however an association was seen between changes in NMOV and amplitude of nystagmus (Pearson correlation coefficient [r]=0.455, P=0.01).

The 2-Rectus Group

This group included 13 male (56.6%) and 10 female (43.5%) patients with mean age of 19.2±11.5 (range 5-48) years. Nystagmus was of motor origin in 12 (52.2%) and sensory origin in 11 (47.8%) cases. Patients were followed for a mean period of 18.7±8.0 (range 6-36) months.

Seventeen patients (73.9%) underwent bimedial recession (BMR) with mean recess of 12.3±0.6 mm and six patients underwent bilateral recession (BLR) with mean recess of 15.3±0.8 mm from the limbus. Each millimeter of BMR recess resulted in 1.9±.9 PD correction of esotropia and each one millimeter of BLR
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recess led to 4.5±6.9 PD correction of exotropia. Mean tropia decreased from 45.9±15.7 PD preoperatively to 3.5±6.1PD in the BMR recess group (P<0.001) and from 53.3±15.4 PD to 8.3±6.8 PD in the BLR recess subgroup (P<0.001) at final follow-up. None of the subgroups developed motility restriction postoperatively.

Mean FBOV and mean FMOV increased significantly after correction of nystagmus in this group but there was no significant correlation between changes in FBOV with the amount of recession in the BMR (P=0.2) or BLR (P=1) subgroups. There was also a non-significant increase in mean NBOV and NMOV postoperatively (table 2). Quality of vision improved in 14 (60.9%) and remained unchanged in 9 (39.1%) cases. There was no significant correlation between improvement in visual quality and the increase in FBOV or NBOV.

Mean SE had a mild myopic change postoperatively which was statistically significant. Mean astigmatism also had a small but non-significant myopic change postoperatively (table 2). The axis of astigmatism remained unchanged in 20 eyes (43.5%), whereas showed a shift to WTR in 18 (39.1%) and a shift to ATR in 8 (17.4%) eyes (P=0.07).

Although non-significant, the frequency, amplitude and speed of nystagmus increased postoperatively (table 3). There was no correlation between improvement in FBOV and NBOV with changes in electronystagmographic indices, however the increase in NMOV had negative correlation with the speed of nystagmus (r=-0.453, P=0.03) and improvement in FMOV had negative correlation with the frequency (r=-0.83, P=0.04) and speed (r=-0.812, P=0.05) of nystagmus.

DISCUSSION

Congenital nystagmus is bilateral, conjugate, horizontal, pendular and jerky on side gazes. The coexistence of slight rotary movements is also common. Involuntary head jerk, although common, has no effect on visual acuity and does not accompany oscilopsia, a helpful feature distinguishing congenital nystagmus from acquired types. Congenital nystagmus is accentuated during focusing on an object, diminishes with accommodation and disappears with sleep. Even when there is no attenuation of the nystagmus during accommodation, better near vision is found for unknown reasons. Anxiety increases the severity of nystagmus. Visual acuity is often decreased but the degree of visual loss depends on the type of associated sensory disorders. A patient with nystagmus often takes
an abnormal head position to maintain the eyes in a neutral point (the point of minimum nystagmus amplitude) to improve vision. Abnormal head posture may not be constant and is usually most obvious when there is need for precise vision, especially for far vision tasks.1

Nystagmus is probably a sensory neurologic disorder. In approximately 90% of cases, an underlying sensory deprivation exists, the most common of which includes different phenotypes of albinism, optic nerve anomalies (atrophy and hypoplasia), retinal abnormalities (Leber’s congenital amaurosis, achromatopsia, congenital stationary night blindness and aniridia), and congenital cataracts. Approximately 10% of patients with nystagmus have relatively good vision, the so-called idiopathic congenital nystagmus or congenital motor nystagmus.1 In our series, 25 patients (43%) had a sensory disorder including oculocutaneous albinism in 22 cases and aniridia, optic nerve atrophy and high myopia each in one case; the remainder had congenital motor nystagmus. The reason for the lower prevalence of sensory disorders in our series may be the referral nature of our center and non-random sampling.

Despite the similarity of symptoms in different types of nystagmus such as visual loss, oscillopsia, abnormal head posture or torticollis, management strategies are various. The first treatment was introduced by Collburn17 in 1906 who sutured the horizontal rectus muscles to the orbital periosteum in order to reduce muscular contraction thereby decreasing the amplitude of nystagmus. In 1950, Metzger18 used a prism with the apex oriented in the direction of the null point in order to correct abnormal head posture. In 1953, Kestenbaum19 performed recession and resection of the rectus muscles in both eyes to approximate the null point to the midline. In this way, both eyes were displaced in the same direction. In the same year, Anderson20 performed yoke muscle recession in the direction of rotation of the eyes. Both methods resulted in improvement of visual acuity and head posture. In 1945, Goto21 performed resection on the medial rectus muscle of one eye and the lateral rectus muscle of the fellow eye to displace the null point and to correct the abnormal head posture. Blatt and Kruznun22 displaced the rectus muscles in the direction of head rotation to weaken muscle function in their range of action. Arruga23 reported a decrease in the amplitude of nystagmus in a patient who underwent a posterior fixating suture on the medial rectus muscles during retinal surgery. Cuppers24 created exotropia by means of surgery to reduce the amplitude of nystagmus. Bietti25 was the first to report recession of all four rectus muscles 12 mm posterior to the equator. The first work in the surgical correction of vertical nystagmus was reported by Pierse26 who corrected abnormal head posture and improved visual acuity by weakening the superior oblique and inferior rectus muscles. Pratt-Johnson27 recommended symmetrical surgery on all four rectus muscles in patients with nystagmus but no strabismus.

Extensive recession of the horizontal rectus muscles behind the equator decreases the speed of eye movements by changing the axes and length-tension of the muscles. Since both agonist and antagonist muscles are weakened proportionately, the procedure may not result in significant limitation of ductions. Cooper and Sandall28 performed recession and resection procedures on both eyes to correct nystagmus in the fixator eye and deviation in the fellow eye. In 1969, Parks29 modified the innovative Kestenbaum method by performing maximal surgery on the muscle in spite of similar surgery on both eyes. He performed 5 mm recession on the medial rectus and 8 mm resection on the lateral rectus muscles in one eye along with 6 mm resection on the medial rectus and 7 mm recession on the lateral rectus in the fellow eye. This method is called the maximum classic method. Calhan and Harley30 refined this method because of the high incidence of under-correction such that by 40% reinforcement, corresponding amounts of surgery were changed to 11.2, 9.8, 8.4 and 7 mm. They recommended that patients with less than 15° face turn do not need any surgical intervention but the amount of surgery should be aug-
mented by 40% in cases with 30° face turn, and by 60% in subjects with 45° face turn using Parks’ method. Taylor31 reported acceptable results from non-symmetrical recession and resection of medial and lateral rectus muscles. He performed recession of the lateral rectus muscle in the direction of the slow phase of nystagmus about 8-9 mm in one eye along with recession of the medical rectus muscle of the fellow eye about 6 mm together with about 6 mm resection of the two remaining muscles.

In all the above-mentioned studies and many other reports in recent years, the amplitude of nystagmus was reduced but visual acuity remained unchanged in some or improved objectively or subjectively in others. Abnormal head posture was corrected in 76-100% of cases during a follow-up period ranging from 3 weeks to 6 years. No study reported marked limitation of eye movements such that the maximum reported limitation was less than 50% restriction in duction which was gradually relieved.2-31 Graf et al32 retrospectively studied 34 patients in order to determine the amount of surgery in the Kestenbaum method and reported 67% decrease in abnormal head posture and found that surgery has an efficacy of 0.8° per each millimeter of surgery on both eyes in the short term and 1.5° per each millimeter of surgery in the long term. They recommended surgery on muscles of each eye equivalent to two-thirds of the amount of face turn. Our previous study revealed that weakening all four horizontal rectus muscles seems to be an appropriate way for decreasing the amplitude of nystagmus and improving visual acuity but is not a reliable method for correction of abnormal head posture.16

In the present study, FMOV improved in all three groups and FBOV improved in the 2-rectus group. In the latter group, because of the large amount of tropia (>30 PD), surgery was planned to simultaneously correct the tropia and nystagmus using maximal recession of both correlated muscles (recession of both lateral recti for exotropia and recession of both medial recti for esotropia). Visual improvement in these cases, especially improvement in far vision, was probably due to the improvement in binocular vision resulting from alignment of the visual axes of the eyes. In the 4-rectus group, although FMOV significantly improved after surgery, improvement in NBOV and FBOV was not statistically significant. Recent studies on tenotomy of rectus muscles indicate that tenotomy alone without muscle transposition can improve visual acuity32-37 which is consistent with our results. The KA group in which surgery was performed on the basis of abnormal head posture including one recession and one resection procedure in either eye, revealed significant improvement only in FMOV which might be due to correction of head position.

All three groups in the present study revealed a decrease in the speed, frequency and amplitude of nystagmus; however, only the decrease in amplitude and speed of oscillations was significant only in the 4-rectus group probably due to the larger number of cases. It is notable that the frequency of nystagmus did not show any significant change in any group. We believe that frequency is the constant component of nystagmus and cannot be changed by muscle surgery.

We noticed a mild myopic shift in the 4-rectus and 2-rectus groups and a hyperopic shift in the KA group. Previous studies in this regard showed that most changes in refractive error following strabismus surgery are transient and non-significant, and that recession has greater effect in this regard as compared to resection. Recession of all four rectus muscles can decrease the total refractive power of the cornea by flattening it. This also holds true for the 2-rectus group in which two muscles on the same meridian are recessed. Some cases of myopic shift have also been reported, however further research is needed to draw definite conclusions.38-42

The study by Hertle et al33 is very similar to our study; they demonstrated that no correlation exists between visual outcomes and electrophysiologic findings, which is consistent with our results. We interestingly found that frequency is the constant property of nystag-
mus such that even if other electrophysiologic indices are affected, frequency is not significantly changed.

In conclusion, surgery on extraocular muscles in congenital nystagmus can improve visual acuity in most patients. With an appropriate choice of procedure, one can correct the abnormal head posture and tropia as well. The correlation between improvement in clinical and electroneystagmographic features is not clear.

REFERENCES