Improved visual acuity and recognition time in nystagmus patients following four-muscle recession or Kestenbaum-Anderson procedures

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PURPOSE

Many patients describe more rapid recognition of objects after surgical procedures for nystagmus; however, this “recognition time” is not reflected in the parameters typically studied in these patients. The purpose of this study is to assess the effect of nystagmus surgery on visual acuity and recognition time.

METHODS

In this prospective, interventional, comparative case series, patients with nystagmus were divided into two groups. Group A (n = 13) underwent four-muscle retroequatorial recession; group B (n = 8) underwent the Kestenbaum-Anderson procedure. Visual acuity, binocularity, and recognition time were assessed before and after surgery. Recognition time was measured in a routine examination setting using specially designed software that controlled the time of appearance of optotypes in 0.1 second increments.

RESULTS

A total of 21 patients were enrolled. The entire group experienced significant postoperative improvement in visual acuity (P = 0.002) and recognition time (P = 0.005). The mean improvement in recognition time was 0.3 seconds at maximum preoperative visual acuity level. A trend toward more improvement in group A than in group B was not statistically significant.

CONCLUSIONS

Both the four-muscle recession and the Kestenbaum-Anderson procedures resulted in a 1- to 2-line improvement in visual acuity and a 0.3 second improvement in optotype recognition time. (J AAPOS 2012;16:36-40)

Surgical treatment of infantile nystagmus syndrome with or without ocular and oculocutaneous albinism includes the Kestenbaum-Anderson procedure, four-muscle recession, four-muscle tenotomy, and combinations of these procedures. The results of the procedures have evaluated subjective visual function (Snellen visual acuity and patient subjective change), objective visual function (eg, amplitude of nystagmus and foveation time), and calculated nystagmus acuity function (NAF) and expanded nystagmus acuity function (NAFX). The evaluations have generally shown positive results from surgery for nystagmus; however, these assessments often do not simulate natural viewing conditions.

Subjects and Methods

Recruitment and work were conducted in accordance with the Declaration of Helsinki. Institutional Review Board approval was granted before the commencement of the study. Written informed consent was obtained for all patients or their guardians.

All patients were at least 5 years old and able to cooperate for visual acuity testing, timed visual acuity testing, and binocularity tests. Patients with and without abnormal head posture were included but assigned to different surgical procedures as mentioned below. Patients with horizontal and/or vertical nystagmus were included. The concomitant presence of strabismus was not an exclusion criterion. Patients whose nystagmus was clinically observed to have disconjugacy, or a fast-phase beating toward the fixating eye, were excluded—the intent was to exclude *spasmus nutans* and manifest latent nystagmus. Additionally, patients with acquired nystagmus and those with sensory deficits other than albinism were excluded, as were patients with visual acuities...
<20/400 (typically with a sensory deficit) and patients who had undergone previous ocular surgery, whether an extraocular muscle surgery or an intraocular procedure.

For pre- and postoperative assessment, the same clinical examination protocol was applied preoperatively and 4 months postoperatively. Patients were assessed on two separate days at least a week apart. All tests were repeated on both examination days to ensure consistency. Initially, a complete ocular examination including a cycloplegic refraction was performed. Visual acuity testing was performed using the tumbling E charts with single optotype using the logMAR scale, using full spectacle correction nonmonocularly and binocularly. Binocularity was assessed using Bagolini glasses, the Worth 4-dot test (Western Ophthalmic, Lynwood, WA) for distance, and Lang stereotest. Head posture was evaluated in every case during binocular visual acuity examination. For cases in which a stable abnormal head posture was detected and confirmed by history, the angle was measured using a goniometer.

Recognition Time Testing
Recognition time was measured in all cases using specially designed software installed on the computer used for regular visual acuity testing. The viewing screen was 4.5 meters from the examination chair, and the tumbling E optotype size was calibrated for the examination distance. Patients were allowed to assume the abnormal head posture for maximum visual acuity and were tested with both eyes open and full spectacle correction. The test was demonstrated using large optotypes until the patient clearly understood the task. The test began with optotypes 2 logMAR lines above the maximum visual acuity of the patient. The target was initially presented with three different optotypes presented at 0.1 second intervals. A response was considered positive if two of three were identified correctly. If the response was negative, the same size optotype was presented for 0.2 seconds duration. The duration was increased by 0.1 second increments until a positive response was obtained. In the second stage of the test, the size of the optotype was increased one logMAR level, that is, one logMAR line less than maximum visual acuity, and testing started at a 0.1 second duration following the same increment protocol. The third and final stage was done using the same protocol at the maximum visual acuity level. After testing at the three levels of logMAR acuity, the patient was allowed to rest for about 20 minutes and the entire procedure was repeated to ensure consistency. The recognition time protocol was repeated on a separate examination day at least 1 week after the first examination and the best results were used.

Procedure Selection
The patients were divided into two groups, independent of recognition time testing results. Group A (four-muscle recession group) included patients with no abnormal head posture. Those that had no strabismus underwent a retroequatorial recession of all four horizontal rectus muscles. The medial rectus muscle was recessed 9 mm from insertion and the lateral rectus muscle was recessed 12 mm from insertion in both eyes. For cases with strabismus and no abnormal head posture, the surgical numbers were modified in an attempt to correct the strabismus. Group B (Kestenbaum-Andersen procedure group) included patients with abnormal head posture. Recession of two rectus muscles was combined with resection of the two antagonist rectus muscles.

Statistical Analysis
Improvements in visual acuity and recognition time were calculated as value after surgery and value before surgery. Statistical analyses were performed using SPSS software version 18.0 (IBM Corporation, Armonk, NY), and statistical significance was set at a 95% confidence level. Testing for normal distribution of data was performed using One-sample Kolmogorov-Smirnov test. The differences between groups A and B were tested using independent samples t test for quantitative variables and the Fisher exact test for categorical variables. A paired sample t test was used to test for significance of change before and after surgery. On further classifications of groups A and B, nonparametric tests were used: Mann-Whitney test to compare values between the two groups and Wilcoxon signed-rank test to compare values before and after surgery.

Results
A total of 21 patients had surgery, with 13 in group A and 8 in group B. There were no statistically significant differences between group A and group B with regard to age (P = 0.47) or sex (P = 0.4) (Table 1). Four group A patients had strabismus and hence the numbers for their four-muscle recession were adjusted to correct the strabismus (Table 2). Case details for all group A patients are provided in e-Supplement 1 (available at www.jaapos.org). No group B patients had strabismus. The muscle operated on and amount of surgery were based on the direction and degree of abnormal head posture, as shown in Table 2. The degree of abnormal head posture for all group B patients is provided in e-Supplement 2 (available at www.jaapos.org). Group A was further subdivided into two groups: group A1 (nine patients), with no sensory deficits, and group A2 (four patients), with oculocutaneous albinism.

Best-corrected visual acuity was significantly better in group A patients after surgery (Table 3). Although visual acuity also showed a tendency toward improvement after surgery in group B patients, the change was not statistically significant. Although the improvement of visual acuity in group B patients appeared less notable than in group A patients, this difference also was not significant.

Recognition time improved significantly after surgery in group A patients, when tested using optotypes representing both the best-corrected preoperative visual acuity optotype and the one logMAR line less than the maximum preoperative visual acuity (Table 4). There was no significant improvement in group B patients. When group A patients were subdivided into groups A1 and A2, only the improvement in group A1 was statistically significant (Table 4). Binocularity did not show any significant change after surgery.
Table 1. Distribution of patients by age and sex

<table>
<thead>
<tr>
<th></th>
<th>Group A</th>
<th>Group B</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>13</td>
<td>8</td>
<td>21</td>
</tr>
<tr>
<td>Males</td>
<td>7</td>
<td>6</td>
<td>13</td>
</tr>
<tr>
<td>Average age (years) ± SD (range)</td>
<td>16.23 ± 5.46 (8-25)</td>
<td>14.25 ± 6.88 (7-22)</td>
<td>15.48 ± 5.95 (7-25)</td>
</tr>
</tbody>
</table>

Table 2. Surgical groups for patients

<table>
<thead>
<tr>
<th>Alignment</th>
<th>Angle (PD)</th>
<th>No. patients</th>
<th>Surgery</th>
<th>AHP direction</th>
<th>Degree range</th>
<th>No. patients</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>No strabism</td>
<td>0</td>
<td>8</td>
<td>MR recess 9 LR recess 12</td>
<td>Right face turn</td>
<td>20-30</td>
<td>2</td>
<td>e-Supplement 3a</td>
</tr>
<tr>
<td>ET</td>
<td>20-30</td>
<td>3</td>
<td>MR recess 6 LR recess 12</td>
<td>Left face turn</td>
<td>20-40</td>
<td>4</td>
<td>e-Supplement 3</td>
</tr>
<tr>
<td>XT</td>
<td>40</td>
<td>1</td>
<td>MR recess 5 LR recess 12</td>
<td>Chin up</td>
<td>25</td>
<td>1</td>
<td>IR recess 5 SR resect 5</td>
</tr>
<tr>
<td>XT</td>
<td>20-30</td>
<td>3</td>
<td>MR recess 6 LR recess 12</td>
<td>Chin down</td>
<td>30</td>
<td>1</td>
<td>SR recess 8 IR resect 5</td>
</tr>
</tbody>
</table>

AHP, abnormal head posture; IR, inferior rectus muscle; LR, lateral rectus muscle; MR, medial rectus muscle; PD, prism diopters; recess, recession; resect, resection; SR, superior rectus muscle.

*Available at jaapos.org. Surgical numbers as described by Wright et al.12

Table 3. Effect of surgery on best-corrected visual acuity

<table>
<thead>
<tr>
<th></th>
<th>Pre-op BCVA</th>
<th>Post-op BCVA</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Group A</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Group A1</td>
<td>0.52 ± 0.18</td>
<td>0.45 ± 0.20</td>
<td>0.009</td>
</tr>
<tr>
<td>Group A2 (OCA)</td>
<td>0.4a</td>
<td>0.3a</td>
<td>0.02</td>
</tr>
<tr>
<td>Group B</td>
<td>0.25 ± 0.18</td>
<td>0.20 ± 0.19</td>
<td>0.11</td>
</tr>
<tr>
<td>Total</td>
<td>0.42 ± 0.22</td>
<td>0.36 ± 0.23</td>
<td>0.002</td>
</tr>
</tbody>
</table>

BCVA, best-corrected visual acuity; OCA, oculocutaneous albinism; Post-op, postoperative; Pre-op, preoperative.

*Median used for calculations.

Improvement in best-corrected visual acuity for both groups A and B ranged from −0.2 to 0 logMAR, yet the median improvement in best-corrected visual acuity for group A (−0.1) was higher than group B (0), which was not statistically significant (P = 0.595). Also, improvement in recognition time for group A ranged from −0.3 to 0.8 seconds, whereas that of group B ranged from −0.7 to 1.4 seconds. The average improvement in recognition time for group A (0.3) was higher than group B (0.2). This was not statistically significant (P = 0.645).

Discussion

Assessment of visual function and effects of treatment modalities in nystagmus patients may require more than standard, static visual acuity measures. Early reports identified foveation time (time of the ocular oscillation cycle in which the image is close to fovea and the eye is moving slow enough to allow useful vision) as the primary determinant for visual function in nystagmus patients. Thus, treatment of nystagmus often aims at increasing the foveation quality. The NAF, calculated from nystagmus waveforms, was thought to correlate closely with visual performance.10 The NAFX was developed for subjects unable to control fixation within the foveation window of the NAF.11 Yang and colleagues16 tested the effect of eccentric gaze and time restriction on visual acuity in infantile nystagmus syndrome patients and concluded that time-restricted visual acuity measurement may be a useful measure for visual function in infantile nystagmus syndrome. Hertle and colleagues17 studied latency of dynamic and gaze-dependent optotype recognition in infantile nystagmus syndrome patients and showed that latencies differed between infantile nystagmus syndrome patients and controls and concluded that these latencies can be a useful measure of visual function in nystagmus patients. One of the earliest reports on objective benefits of four-muscle recession was published in 1995 by Egbert and colleagues,18 who described a patient with fusional maldevelopment nystagmus and oculocutaneous albinism. Using magnetic scleral search coils, they found that surgery led to an increase in the duration of the low-velocity portion of the nystagmus. Sprunger and colleagues19 assessed the effect of four-muscle recession on recognition time in three patients and concluded that surgery decreased the time necessary to recognize optotypes and therefore improved visual function. The “slow to see” phenomenon was also studied by Wang and colleagues20,21 using a behavioral ocular motor system model. They investigated the latency of the initial reflexive saccade and latency to target acquisition and simulations predicted lengthy target acquisition times.20,21 Recognition time used in the present study simulated natural viewing conditions and allowed the clinician and patient to understand better and appreciate the effect of surgery.

Kestenbaum-Andersen procedures were shown to correct abnormal head posture associated with nystagmus as early as the 1950s.1,2 Decades later, investigations revealed beneficial effects on visual function by broadening the null region and causing overall reduction in nystagmus.
intensity in all positions of gaze. Similar improvement in visual acuity has been demonstrated for procedures whose primary outcome goal was to improve vertical or horizontal abnormal head posture. Kumar and colleagues have demonstrated significantly improved visual acuity from logMAR of 0.42 to 0.33 ($P = 0.002$) after Kestenbaum-Andersen procedures.

Our reported improvement may not reflect the effect of surgery on overall visual function because visual acuity and recognition time were evaluated independently rather than factored together. Calculation of recognition time improvement was done by subtracting postoperative from preoperative recognition time measured at the same level of visual acuity. For those patients who showed an improvement in visual acuity, the overall visual function may have improved, yet this was not calculated because of a lack of a statistical function for such assessment. Hence a small improvement in visual acuity, combined with a small improvement in recognition time, may indeed improve visual quality in a way that cannot be statistically evaluated.

Four-muscle recession surgery, introduced in the 1950s, went out of favor for a period of time before being revived again by Von Noorden and colleagues and Helveston and colleagues in the early 1990s. Datta and colleagues reported a decrease in nystagmus amplitude and improvement in visual acuity in nine patients that underwent the procedure, and Erbagci and colleagues reported that five of seven patients experienced improved visual acuity and head posture (71.4%). Boyle and colleagues reviewed a relatively large series of 18 patients who underwent four-muscle recession and found an improvement of one line of Snellen acuity in nine patients, with the remainder describing only minor improvement. They concluded that the change in acuity is of limited objective benefit as indicated by their patient questionnaire, which returned 8 satisfied patients of 12. Although the design of the questionnaire was a limitation of this study, it shed light on another outcome measure of this surgery, that is, its effect on daily life.

The tenotomy and reattachment procedure was first described on canines to have a nystagmus damping effect. Further studies on humans showed that tenotomy improved visual function as measured by the NAFX. Both the four-muscle recession procedure and the Kestenbaum-Andersen procedure involve tenotomy; however, the strong resection in the Kestenbaum-Andersen procedure may cause this subset of patients to respond differently. Results with our group B patients might be expected to differ from those of group A because the former group’s lower preoperative visual acuity may have allowed for greater postoperative improvement; additionally, assessment for group B was performed with both eyes open, allowing patients to assume a head posture conducive to maximum vision performance.

The presence or absence of ocular or oculocutaneous albinism does not affect diagnosis of infantile nystagmus syndrome because it is based mainly on waveform. Extraocular muscle surgery on patients with oculocutaneous albinism and nystagmus has produced favorable results, comparable to those of infantile nystagmus syndrome without any sensory deficit. In the present study, the improvement of visual acuity and recognition time for patients with albinism was not statistically significant; however, our clinical impression was that the satisfaction rate was similar to patients with no sensory deficit. Less improvement is expected in cases with oculocutaneous albinism than in those without sensory deficit, yet the results of this study are not sufficiently definitive to recommend either performing or not performing surgery in this subset of patients. Further work with a larger number of albinism subjects may reveal more about this group.

Limitations of our study include the small number of patients, lack of masking, absence of a control group, and the possibility that patients perform better with repeated testing. The method of testing was standardized to try to avoid examiner bias. Further work on these patients using eye movement recordings in concert with recognition time testing will shed more light on visual improvement following nystagmus surgery.

Table 4. Effect of surgery on optotype recognition time

<table>
<thead>
<tr>
<th>Group</th>
<th>Pre-op RT1</th>
<th>Post-op RT1</th>
<th>P value</th>
<th>Pre-op RT2</th>
<th>Post-op RT2</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>A (4MR)</td>
<td>0.62 ± 0.3</td>
<td>0.31 ± 0.18</td>
<td>0.003</td>
<td>0.31 ± 0.19</td>
<td>0.15 ± 0.52</td>
<td>0.018</td>
</tr>
<tr>
<td>Group A1</td>
<td>0.6a</td>
<td>0.2a</td>
<td>0.01</td>
<td>0.35a</td>
<td>0.1a</td>
<td>0.41</td>
</tr>
<tr>
<td>Group A2 (OCA)</td>
<td>0.4a</td>
<td>0.35a</td>
<td>0.79</td>
<td>0.15a</td>
<td>0.15a</td>
<td>1</td>
</tr>
<tr>
<td>Group B (KAP)</td>
<td>0.81 ± 0.98</td>
<td>0.54 ± 0.57</td>
<td>0.24</td>
<td>0.23 ± 0.95</td>
<td>0.18 ± 0.11</td>
<td>0.41</td>
</tr>
<tr>
<td>Total</td>
<td>0.69 ± 0.63</td>
<td>0.39 ± 0.38</td>
<td>0.005</td>
<td>0.27 ± 0.17</td>
<td>0.16 ± 0.08</td>
<td>0.012</td>
</tr>
</tbody>
</table>

*Post-op, postoperative; Pre-op, preoperative; RT1, recognition time measured at level of preoperative best-corrected visual acuity; RT2, recognition time measured at level of visual acuity one logMAR line less than preoperative best-corrected visual acuity.

aMedian used for calculations.

References

20. Wang ZI, Dell’Osso LF. Being “slow to see” is a dynamic visual function consequence of infantile nystagmus syndrome: model predictions and patient data identify stimulus timing as its cause. Vision Res 2007;47:1530-60.