Spasmus Nutans
A Long-term Follow-up
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Purpose. Nystagmus, head nodding, and anomalous head position are symptoms of spasmus nutans. This disorder appears in early childhood and is thought to be self-limited. However, the visual outcome of patients with spasmus nutans is unclear. The resolution of nystagmus has not been proven with quantitative eye movement recordings. The purpose of this study was to perform long-term follow-up examinations (mean, 5.5 years) of patients with spasmus nutans.

Methods. Ten patients with spasmus nutans were followed up clinically until a mean age of 7 years. Included were quantitative eye and head movement recordings.

Results. At their last examination (mean age, 7.1 years), visual acuity in four patients was 20/20 in both eyes, in five patients it was 20/30 or better in at least one eye, and in one patient it was 20/50 in each eye. Three patients had orthotropia with normal stereo acuity. The remaining patients had esotropia, dissociated vertical deviation, amblyopia, or latent nystagmus. All patients had fine, intermittent asymmetric, pendular nystagmus on eye movement recordings.

Conclusions. Good visual acuity can be expected in patients with spasmus nutans; one third have normal stereo acuity. However, subclinical nystagmus persists until at least 5 to 12 years of age. Invest Ophthalmol Vis Sci. 1995;36:2768–2771.

Spasmus nutans includes asymmetric nystagmus, head nodding, and anomalous head position.1–6 The neural substrate of this disorder remains unknown. It occurs in early childhood and has been reported to be a self-limited disorder.1–5 Previously, one of the essential criteria of the diagnosis of spasmus nutans has been the spontaneous resolution of signs. However, more objective criteria based on eye and head movement recordings are being used to differentiate spasmus nutans from other forms of nystagmus.5–6 Few longitudinal studies have been conducted in patients with spasmus nutans.3,5,6 Norton and Cogan8 observed, in a clinical study, that symptoms in 5 of 20 patients persisted for 3 years or more. Weissman et al5 repeatedly examined seven children with spasmus nutans with quantitative eye movement recordings between 4 and 30 months. One child had resolution of nystagmus at 12 months of age. Another child showed no ocular oscillations on clinical examination at 17 months of age. Persistence of nystagmus, however, was evident on eye movement recordings. In our previous study,6 none of the patients examined between 3 months and 8 years of age with eye and head movement recordings had resolution of nystagmus at the last examination. Therefore, it is unclear whether spasmus nutans is, as thought, self-limited or whether subtle abnormalities persist even in patients whose routine evaluations showed resolution of clinical signs. The visual acuity and binocularity of patients with a history of spasmus nutans are unknown.

The aim of this study was to examine visual functions of former patients with spasmus nutans and to analyze whether symptoms of spasmus nutans disappear after infancy. Using clinical examination and quantitative eye and head movement recordings, we followed 10 patients with spasmus nutans up to a mean age of 7 years.

PATIENTS AND METHODS. Research followed the tenets of the Declaration of Helsinki. Parents of all subjects gave informed consent, and the study was approved by the Institutional Review Board Committee of Wills Eye Hospital. Included in this study were 10 patients with spasmus nutans who were first seen between 6 months and 4 years of age at the Foerderer Eye Movement Center, at Wills Eye Hospital. They underwent ophthalmic examination and quantitative eye and head movement recordings. The following criteria of spasmus nutans were met at their first examination: fine, rapid, pendular, dissociated (different nystagmus amplitude of the two eyes and/or right and left eye oscillating out of phase) nystagmus; head nodding (abnormal horizontal, vertical, or oblique head movements of 3 to 5 Hz suppressing the nystagmus); and normal results on computed tomography or magnetic resonance imaging. Several follow-up examinations (mean 5.8 examinations) were performed on all patients until they were 5 to 12 years (mean age, 7.1 years) (see Table 1). A full ophthalmologic examination, as well as eye and head movement recordings, were obtained at each examination. Snellen visual acuity, alternating prism cover test, analysis of eye motility, and stereoacuity by Lang, Titmus
TABLE 1. Patient Characteristics

<table>
<thead>
<tr>
<th>Patient Number</th>
<th>Age at First Examination (years)</th>
<th>Number of Eye Movement Recordings</th>
<th>Age (years)</th>
<th>Ocular Alignment</th>
<th>Visual Acuity</th>
<th>Nystagmus</th>
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<tbody>
<tr>
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<td></td>
<td></td>
<td></td>
<td>Right Eye</td>
<td>Latent/Manifest</td>
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<tr>
<td>1</td>
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<td>2</td>
<td>6</td>
<td>ET</td>
<td>20/40</td>
<td>7</td>
</tr>
<tr>
<td>2</td>
<td>1</td>
<td>9</td>
<td>6</td>
<td>ET DVD</td>
<td>20/30 20/30</td>
<td>8</td>
</tr>
<tr>
<td>3</td>
<td>0.9</td>
<td>8</td>
<td>8</td>
<td>A-pattern ET DVD</td>
<td>20/30 20/40</td>
<td>5</td>
</tr>
<tr>
<td>4</td>
<td>2</td>
<td>8</td>
<td>7</td>
<td>ET</td>
<td>20/30 20/50</td>
<td>10</td>
</tr>
<tr>
<td>5</td>
<td>0.6</td>
<td>5</td>
<td>6</td>
<td>ET DVD</td>
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<td>10</td>
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<tr>
<td>6</td>
<td>1</td>
<td>7</td>
<td>8.6</td>
<td>ET DVD</td>
<td>20/30 20/30</td>
<td>8</td>
</tr>
<tr>
<td>7</td>
<td>1.1</td>
<td>6</td>
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<td>Orthotropia</td>
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<td>15</td>
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<tr>
<td>8</td>
<td>0.8</td>
<td>5</td>
<td>5.7</td>
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<tr>
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<td>4</td>
<td>4</td>
<td>12</td>
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<tr>
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<td>4</td>
<td>7</td>
<td>Orthotropia</td>
<td>20/20 20/20</td>
<td>5</td>
</tr>
</tbody>
</table>

DVD = dissociated vertical deviation; ET = esotropia.

Analysis of the recording gave oscillation frequency, phase relationships, and head movements from a period of 2 to 5 minutes. The patient was considered to have nystagmus when at least three consecutive eye oscillations, with amplitudes of at least 0.5°, occurred more than five times on eye movement recordings. At less than 0.5°, it was difficult to distinguish between nystagmus and noise. Therefore, smaller oscillations were not considered nystagmus.

RESULTS. Clinical Findings. At the last examination, 7 of 10 patients had associated ocular findings such as esotropia, dissociated vertical deviation, or amblyopia. Three patients had orthotropia with normal stereo acuity. Five patients had, in addition, manifest latent nystagmus. Visual acuities ranged between 20/20 (both eyes in four patients) and 20/50. Nine patients had visual acuity of 20/30 or better in at least one eye. Reduced visual acuity was noted in the five patients with manifest latent nystagmus. In addition, visual acuity was reduced to 20/50 in patient 4 without latent nystagmus on eye movement recordings. In this 7-year-old patient, small asymmetric pendular nystagmus was detected by clinical examination. No nystagmus was seen clinically in the remaining nine patients. All patients with orthotropia had visual acuity of 20/20 in both eyes. Four patients had been treated with patching therapy for amblyopia.

Eye and Head Movement Recordings. At the last examination in all 10 patients, EOG showed fine, intermittent, asymmetric horizontal and/or vertical pendular nystagmus still appropriate to the diagnosis of spasmus nutans. Nystagmus amplitude, however, was clearly smaller than it was at the first examination and ranged between 0.5° and 3° (mean 1.2°). The
nystagmus frequency was between 5 and 15 Hz. Five of the six patients with esotropia and dissociated vertical deviation displayed typical latent nystagmus beating in the direction of the dominant or uncovered eye with decreasing exponential velocity of the slow phase. Three patients displayed intermittent head nodding on recordings, with an amplitude between 2° and 4° at a frequency between 3 and 5 Hz.

Figure 1A shows an example of the horizontal and vertical eye movement recordings of patient 9 at age 8 years during 10° right horizontal saccadic eye movements. Fine, intermittent nystagmus is seen on the

FIGURE 1. Original electro-oculogram recordings of eye and head positions of patient 9 at 8 years of age reproduced from paper tracings (A). Horizontal eye and target positions, generated from magnetic tape, of the same patient at 12 years of age during horizontal saccadic eye movements of 10° (B). (a) = right eye horizontal position; (b) = right eye vertical position; (c) = left eye horizontal position; (d) = left eye vertical position; (e) = horizontal target position; (f) = horizontal head position; (g) = vertical head position. On horizontal tracings, upward movements depict movements to the right, and downward movements depict movements to the left. On vertical tracings, upward movements depict movements up, and downward movements depict movements down. Arrow indicates primary position.
vertical eye movement tracing of the right eye and on the horizontal eye movement tracing of the left eye.

Figure 1B shows the horizontal eye movement recordings of patient 9 at 12 years of age during 10° left horizontal saccadic eye movements. Fine dissociated pendular nystagmus can be observed in both eyes.

DISCUSSION. In our study, all patients with spasmus nutans had, at their last examination, Snellen visual acuity of 20/50 or better. Nine of the 10 patients had visual acuity of 20/30 or better in at least one eye. Therefore, good visual acuity can be predicted in patients with spasmus nutans. Reduced visual acuity appears to be caused by associated manifest latent nystagmus and amblyopia. Approximately two thirds of the patients had symptoms of congenital squint syndrome, such as esotropia, dissociated vertical deviation, amblyopia, and latent nystagmus, whereas one third had orthotropia with normal stereo function. It is difficult to conceive that patients with asymmetric nystagmus can develop normal stereovision. However, it has been demonstrated that patients with spasmus nutans can suppress their nystagmus by head nodding.8,9 Good visual acuity and stereovision may be developed during phases of compensatory head nodding, while both eyes are stabilized by a normal vestibulo-ocular reflex.

In our study, all patients with spasmus nutans displayed persistent nystagmus on EOG recordings at the last examination at a mean age of 7 years. These findings suggest that many cases described clinically as resolved may have merely improved to reach a subclinical level, and are in agreement with the findings in a patient observed by Weissman et al.9 Therefore, spasmus nutans does not seem to be a self-limited disease in infancy, as previously thought.1-9 We observed bursts of fine pendular nystagmus in several adult patients with latent nystagmus. In these patients, a history of spasmus nutans in infancy must be considered.

In conclusion, good visual acuity can be expected in patients with spasmus nutans. Orthotropia and normal stereovision will develop in approximately one third of the patients. However, we observed subclinical fine nystagmus in all patients until approximately 5 to 12 years of age. Therefore, although associated with a good prognosis, spasmus nutans, contrary to its usual characterization, is not self-limited in infancy. Subclinical eye movement abnormalities persist.

Key Words

electro-oculograms, head nodding, nystagmus, spasmus nutans, stereo-acuity

References