Spasmus Nutans
A Quantitative Prospective Study

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- Spasmus nutans includes ocular oscillations, head nodding, and anomalous head positions. No quantitative longitudinal study verifying the natural history of this self-limited condition has appeared in the literature. Using infrared oculography, we prospectively examined the eye movements of otherwise neurologically normal infants in whom a diagnosis of spasmus nutans had been made. At this writing the ocular oscillations and head movements in two of the children were clinically absent, thereby confirming the diagnosis. The ocular movements are characterized by a phase difference between the oscillations of each eye that varies both during one recording session and during development. The dissociated, pendular nystagmus consists of high-frequency oscillations that may be disconjugate, conjugate, or purely unioocular.

Nystagmus in infancy may be due to a variety of disorders. The differential diagnosis includes "idiopathic" congenital nystagmus, latent/manifest latent nystagmus, nystagmus in association with optic nerve glioma, and spasmus nutans. Once optic glioma is excluded, spasmus nutans may be suspected from anomalous head movement and disconjugate oscillations, but, prior to this study, only follow-up could confirm whether a child had congenital nystagmus or spasmus nutans.

Few reports present well-documented recordings of eye movement in infants and children with a diagnosis of spasmus nutans. In none of the recorded cases was the diagnosis of spasmus nutans confirmed, since a longitudinal analysis of the nystagmus as well as clinical confirmation were not included in the reports. Previously, one of the essential criteria in documenting spasmus nutans has been the spontaneous resolution of nystagmus and anomalous head positions.

We have prospectively conducted a study of the ocular oscillations in infants with the presumptive diagnosis of spasmus nutans with the objective of identifying pathognomonic features early in its course. A preliminary report of this work was presented at the conference "Adaptive Processes in Visual and Oculomotor Systems." 

RESULTS

Table 1 summarizes the results of the clinical evaluation. The mean age at onset, based on the mother's account, was 5 months, with a range of birth to 14 months. Five of the seven patients exhibited head nodding or tilting in addition to the ocular oscillations. One of the children seen for a follow-up evaluation demonstrated a resolution of his ocular oscillations ten months after onset, thereby confirming the diagnosis of spasmus nutans. This patient had been observed to have abnormal head movements. Another infant showed no ocular oscillations on the videotaped record although there was occasional head nodding. However, persistence of her nystagmus was evident in the eye-movement records. The patient's mother reported an improvement in this infant's nystagmus and head movement.

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An example of a typical eye-movement recording associated with spasmus nutans is shown in the Figure. The record shows both eyes at three very closely spaced times (within seconds) during a single recording session for one patient (patient 3). The pendular oscillations of both eyes varied from in phase to 180° out of phase within just 2 s. The unihcocular portion of the record occurred 90 s before this interval. Unihocular oscillations were observed in five of seven patients.

Table 2 summarizes the nystagmus characteristics. All patients demonstrated a pendular waveform both in the initial and follow-up records. The frequency of the ocular oscillations ranged from 3 to 10 Hz. Most patients showed a range of frequencies within one recording session. All of the patients exhibited an asymmetry of the amplitudes of the oscillations in the two eyes. Three showed intervals of asymmetric oscillations and other intervals of symmetric waveform amplitudes during a single recording session. All patients showed variable phase relationships between the oscillations of each eye. Four patients exhibited phase differences ranging between 0° and 180°. In one patient the phase difference variability was in a narrower range, remaining between 120° and 180°. The results of analysis (mean and SD) of approximately ten measured cycles from each record, as shown in Table 2, underscore this variability. One infant, patient 3, at the first recording at 10 months of age showed only in-phase oscillations. However, at a subsequent recording seven months later the phase relationships between the waveforms in each eye varied from 0° through 180°.

Analysis of the records also included classification into relative conjugacy of the waveforms. The scale used consists of the following: +2, equal amplitude of waveforms between the two eyes and in phase; +1, unequal amplitude and in phase; 0, unihocular oscillations; −1, unequal amplitude and out of phase; and −2, equal amplitude and out of phase. For purposes of this scale, in phase means that there was a phase difference of less than 90° between the eyes; out of phase means the difference was more than 90°. Six of the seven infants showed variable conjugacy during a single recording session. Although conjugacy varied, most infants showed one particular predominant conjugacy relationship, usually −1 or +1. Two infants exhibited little variability of conjugacy. However, one of these infants showed varying conjugacy at a later recording session.

In one patient, the vestibulo-ocular reflex was stimulated by quasisinusoidal passive movement of the infant’s head in the horizontal plane. This manipulation did not suppress his ocular oscillations. Active head movement in relation to the ocular oscillations was not evaluated.

**COMMENT**

Spasmus nutans is a self-limiting disorder of infancy and early childhood that was first fully described by Raudnitz in 1897. The pathogenesis of this entity remains obscure. Over this period, only four longitudinal studies of the clinical course have been reported, to our knowledge. Few quantitative analyses of the ocular oscillations have appeared in the literature. These patients did not have a firm diagnosis of spasmus nutans, as no follow-up was obtained. The present study is the first study to date that both clinically and oculo-phonetically evaluates over a substantial period infants with a diagnosis of spasmus nutans. This investigation also provides the first quantitative documentation of eye movement recordings in a patient whose acquired, dissociated, pendular nystagmus subsequently resolved. Spasmus nutans is a condition that should be differentiated from other diseases of infancy that have significant morbidity and potential mortality.

Several reports have described cases in which infants presented with
pendular and often asymmetric nystagmus initially diagnosed as spasmus nutans. These infants were later found to have optic nerve and intracranial gliomas. An important clinical feature in all of these patients was the presence of optic nerve atrophy. Some of the patients exhibited other associated clinical findings, such as diencephalic syndrome and evidence of increased intracranial pressure. There is also one report of an incidence.

Table 2.—Characteristics of Spasmus Nutans

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Date of Study</th>
<th>Frequency, Hz</th>
<th>Relative Amplitude</th>
<th>Phase Relations</th>
<th>Conjugacy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1/83</td>
<td>3</td>
<td>R &gt; L; R = L</td>
<td>Range, 0-180; m, 135.5; SD, 52.9</td>
<td>-1, 0, -2</td>
</tr>
<tr>
<td>5/85</td>
<td>3-5</td>
<td>R &gt; L; R = L</td>
<td>Range, 0-180; m, 78.7; SD, 61</td>
<td>+1 &gt; 0, -1, -2</td>
<td>+2, +2</td>
</tr>
<tr>
<td>2</td>
<td>1/84</td>
<td>4-6</td>
<td>R &gt; L; R = L</td>
<td>Range, 0-180; m, 129.7; SD, 73.4</td>
<td>-1, -2, +1</td>
</tr>
<tr>
<td>5/86†</td>
<td>...</td>
<td>...</td>
<td>Range, 0</td>
<td>+1</td>
<td>+2, 0</td>
</tr>
<tr>
<td>3</td>
<td>10/84</td>
<td>7-9</td>
<td>R &gt; L</td>
<td>Range, 0</td>
<td>+1</td>
</tr>
<tr>
<td>6/85</td>
<td>6-7</td>
<td>L &gt; R</td>
<td>Range, 0-180; m, 89.6; SD, 70.5</td>
<td>+1, -1</td>
<td></td>
</tr>
<tr>
<td>3/86</td>
<td>5-6</td>
<td>L &gt; R</td>
<td>Range, 35-160; m, 112; SD, 54.5</td>
<td>0, +1</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>2/85</td>
<td>6</td>
<td>L &gt; R</td>
<td>Range, 120-180; m, 142.1; SD, 22.2</td>
<td>-1</td>
</tr>
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<td>5</td>
<td>4/85</td>
<td>5-6</td>
<td>L &gt; R</td>
<td>Range, 0-180; m, 104.3; SD, 63.7</td>
<td>-1 &gt; +1</td>
</tr>
<tr>
<td>10/85</td>
<td>9</td>
<td>L &gt; R</td>
<td>Range, 15-180; m, 108.9; SD, 72.4</td>
<td>+1, +2</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>1/86</td>
<td>5-8</td>
<td>R &gt; L</td>
<td>Range, 0-45; m, 9; SD, 18</td>
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<tr>
<td>3/86</td>
<td>8-10</td>
<td>R &gt; L</td>
<td>Range, 0-90; m, 32.1; SD, 39.6</td>
<td>+1, 0</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>11/85</td>
<td>8-9</td>
<td>L &gt; R</td>
<td>Range, 90-180; m, 166.2; SD, 29.5</td>
<td>0 &gt; -2, +2</td>
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<tr>
<td>2/86</td>
<td>8-9</td>
<td>L &gt; R</td>
<td>Range, 0-90; m, 36; SD, 33.7</td>
<td>0, +1</td>
<td></td>
</tr>
</tbody>
</table>

All waveforms were pendular. The conjugacy scale is as follows: +2 indicates equal amplitudes, in phase; +1, unequal amplitudes, in phase; 0, unicocular; -1, unequal amplitudes, out of phase; and -2, equal amplitudes, out of phase. m indicates mean. Amplitudes and conjugacies are listed in order of descending incidence.

†No ocular oscillations were present.

The subsequent clinical course and autopsy findings confirmed the diagnosis of Leigh's subacute necrotizing encephalomyelopathy. Congenital nystagmus is a conjugate oscillation that may be first noted during infancy. The diagnostic distinction between congenital nystagmus, latent/manifest latent nystagmus, and spasmus nutans is important. Congenital and latent/manifest latent nystagmus are associated with life-long impairment of visual acuity; the siblings and children of a patient with congenital nystagmus may be similarly affected. Thus, it is important to differentiate congenital nystagmus from spasmus nutans as early as possible.

All of the patients included in our study had normal results of ophthalmologic and neurologic examinations. Although visual acuity was not formally tested, all parents reported a normal ability of the infant to attend to visual tasks such as locating raisins and other small objects without difficulty. All of the patients with acquired pendular nystagmus described by Gresty et al19 complained of oscillopsia that interfered with their daily activities; it is difficult to determine whether infants with spasmus nutans have this problem. By observation they show no gross impairment in their visual function.

Gresty et al11 described three patients who exhibited a distinct pattern of eye-head coordination. All carried a tentative diagnosis of spasmus nutans; head shaking modified their nystagmus so that vision improved. However, all were older children (more than 3 years of age) and follow-up was not obtained. Suppression of nystagmus occurred with both active and passive head shaking. Gresty et al interpreted this as adaptive suppression of nystagmus as due to vestibular stimulation. In one of our patients (patient 3) we passively rotated the infant's head horizontally and observed no modification of the nystagmus. This patient's nystagmus subsequently resolved. Possibly, the difference in actively as opposed to passively generated head movement may account for the apparent discrepancy. Also, our patient had no clear history of anomalous head movement whereas all of the patients described by Gresty et al1 did.

Analysis of the waveforms disclosed that within a single recording session both the frequency of the oscillations and the interocular phase relationships may vary considerably, even from second to second. In two of our patients, a variability in the relative waveform amplitude was recorded during a single session. The characteristics of this dissociated nystagmus varied not only during one recording session but also in different sessions. It should be noted that at times, or even throughout a recording session, the nystagmus may be conjugate and mimic congenital nystagmus.

In addition to the infant in whom we oculographically documented the resolution of the nystagmus, another infant's nystagmus had clinically resolved, and this finding was recorded in her clinic chart. However, her mother noted that nodding was still intermittently present. Even with careful observation, we were unable to detect abnormal eye or head movements on her follow-up videotape record. However, eye-movement recordings corroborated her mother's history since ocular oscillations were

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Spasmus Nutans—Weisman et al 527
readily apparent. Since our recordings showed that the nystagmus associated with spasmus nutans was still present in infancy and early childhood despite our identification of the characteristics of the resulting nystagmus. Future studies should help elucidate the neural substrate of this disorder. This knowledge will help us use recording for early discrimination between spasmus nutans and other ocular oscillations.

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References