Infantile Esotropia With Nystagmus

A Treatable Cause of Oscillatory Head Movements in Children

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Objective: To document the resolution of oscillatory head movements following surgical realignment of the eyes in children with infantile esotropia and nystagmus.

Method: Retrospective review of 3 children who had infantile esotropia, nystagmus, and unexplained head shaking or head nodding.

Results: Strabismus surgery restored ocular alignment and produced resolution of the head shaking in all patients. In 1 patient, head shaking accompanied recurrence of the esotropia and again resolved following surgical realignment of the eyes.

Conclusions: Head shaking or head nodding can rarely be associated with infantile esotropia and nystagmus. In this syndrome, surgical realignment of the eyes may produce simultaneous resolution of the head oscillations.

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REPEITIVE HEAD SHAKING OR head nodding in infancy can be an ominous clinical sign. Causative factors include spasmus nutans, congenital nystagmus, and neurological lesions (particularly tumors of the third ventricle, producing a bobble-headed doll syndrome). The association of horizontal strabismus with oscillatory head movements has been described in 2 children with intermittent esotropia. We treated 3 children with infantile esotropia, nystagmus, and prominent head oscillations that resolved unexpectedly following surgical realignment of the eyes.

METHODS

Two patients from Arkansas Children's Hospital, Little Rock, and 1 patient from Cedars-Sinai Medical Center are described. Before medical record review, study approval was obtained from the institutional review board of the University of Arkansas for Medical Sciences.

REPORT OF CASES

CASE 1

A 5-month-old Kuwaiti girl was referred for esotropia and head bobbing, which were noted after birth. She was otherwise neurodevelopmentally normal. The results of magnetic resonance imaging of the brain were normal. She was the full-term product of a normal pregnancy, labor, and delivery.

Findings from the physical examination showed prominent episodes of head nodding and a fine, symmetrical, torsional nystagmus during periods of visual attention (a video is available at http://www.archophthalmol.com). During near fixation, she had 65 prism diopters (PD) of comitant esotropia with latent nystagmus and crossed fixation. She displayed brisk abduction saccades with full abduction responses to manual head rotation. She had no associated oblique muscle overaction or dissociated vertical divergence. She had no significant refractive error. The results of optic disc and retinal examinations were normal.

At the age of 5 months, she underwent bimedial recessions (6.5 mm OU). Four days postoperatively, her eyes were straight, her head nodding was markedly diminished, and a subtle nystagmus persisted. Six months postoperatively, she had a small exophoria and bilateral dissociated vertical divergence, and her head nodding and nystagmus had completely resolved. Now, at 7 years of age, she has maintained good ocular alignment with no recurrent head nodding or nystagmus.
CASE 2

A 2-year-old boy with infantile esotropia developed persistent vertical head nodding. He was the full-term product of a normal pregnancy and a labor that was complicated by failure to progress, necessitating cesarean delivery. His birth weight was 2.9 kg. His parents reported that his head nodding began at the age of approximately 4 months and that his esotropia was noted shortly thereafter. His esotropia was variable, ranging from 40 to 70 PD at near. Over time, he developed a manifest latent nystagmus associated with amblyopia of the left eye, which was treated with occlusion therapy. His eyes seemed almost straight at times, but turned in when he became tired. Because of feeding problems, he underwent a neurodevelopmental assessment that showed a mild global developmental delay. An extensive neurodiagnostic evaluation, including a neurological examination, electroencephalography, magnetic resonance imaging, and screening for neurometabolic disease; disclosed no abnormalities.

On our initial examination when he was 2 years of age, he followed optokinetic stimuli and maintained fixation briskly using either eye, with a slight fixation preference for the right eye. Both pupils reacted briskly to light, with no afferent pupillary defect. Versions were comitant, and ductions were full. He had a variable esotropia that measured up to 65 PD. A fine, symmetrical, bilateral, horizontal nystagmus was also detected. Horizontal optokinetic responses had a marked horizontal nasotemporal asymmetry. He turned his head to fixate with his esotropic eye and showed a latent nystagmus. When fixating objects of interest, he developed horizontal elliptical head nodding that subsided when fixation was discontinued. The results of an optic disc and retinal examination were normal. Cycloplegic refraction was +2.25 + 1.00 × 105° OD and +1.25 + 1.00 × 83° OS. The results of optic disc and retinal examinations were normal in both eyes.

Wearing his full cycloplegic refraction and a 3.00 bifocal, his eyes remained straight for several months. Six months later, however, he had developed an esotropia of 10 PD at distance and 18 PD at near, with a latent nystagmus. At the age of 3 years, he was treated with bimedial recessions (4 mm OU) that restored ocular alignment and produced resolution of his head nodding. Wearing his glasses, he was orthotropic at distance and had 12 PD of exophoria at near. During the next year, his esotropia recurred and he developed head shaking with an elliptical component. The results of a magnetic resonance imaging of the brain were again normal. During the next several years, he was treated with glasses and intermittent occlusion of the left eye. His parents noted that he only displayed the head oscillations when wearing his glasses.

At the age of 9 years, he had 20 to 25 PD of intermittent esotropia at distance and 30 PD of intermittent esotropia at near while wearing his full cycloplegic refraction, and showed head nodding when fixating objects of interest. He was treated with bilateral lateral rectus muscle resections (5 mm OU), which restored ocular alignment and again eliminated his head nodding.

CASE 3

A 9-month-old girl with hydrocephalus was examined for esotropia, which was reported to have been present since birth. She had undergone successful shunting for her hydrocephalus but had multiple other congenital anomalies, including Chiari malformation type I, tetralogy of Fallot, enlarged kidneys, vertebral anomalies with scoliosis, an accessory rib, and gastroesophageal reflux requiring placement of a gastroesophageal tube. She was diagnosed as having VATER association (vertebral defects, anal atresia, tracheoesophageal fistula, esophageal atresia, and radial and renal dysplasia). Her parents reported that she frequently shook her head from side to side and had developmental delay for which she was undergoing physical and occupational therapy. She was treated with digoxin, aspirin, and macrodantoin. She was the full-term product of a pregnancy that was complicated by methamphetamine abuse. Macrocephaly necessitated her cesarean delivery; she weighed 3.2 kg at birth.

On examination at the age of 9 months, she responded to optokinetic stimuli using either eye. She maintained fixation with either eye but preferred fixation with the right eye. At near fixation, she had 35 PD of V-pattern esotropia with bilateral, inferior, oblique muscle overaction; dissociated vertical divergence; and a fine, symmetrical, torsional nystagmus. Cycloplegic refraction was +3.50 + 1.50 × 60° OD and +3.50 + 1.50 × 110° OS. The results of optic disc and retinal examinations were normal, except for bilateral retinal extorsion.

She was prescribed her full cycloplegic refraction but refused to wear the glasses. Nevertheless, her parents noted that her eyes did not cross as much as before. Indeed, our examinations at the ages of 14 and 17 months showed a residual fixation preference with the right eye, an intermittent esotropia of 40 PD, and a right hypertropia of 10 PD (attributable to inferior oblique overaction) at near fixation. She returned at the age of 19 months, following a shunt revision. Wearing her full cycloplegic refraction, she had 25 PD of constant esotropia at near fixation, with a fine torsional nystagmus. By the age of 2 years, her esotropia had increased to 35 to 40 PD while wearing her glasses. She was treated with bimedial recessions (5 mm OU) and bilateral, inferior, oblique muscle resections (14 mm OU). Four days postoperatively, no residual head shaking was noted, and the parents reported that the head shaking was gone the day after surgery. Two years later, she had a well-controlled intermittent esotropia of 15 to 20 PD, no nystagmus, and no recurrent head shaking.

In 1990, Rubin and Slavin described a neurologically normal infant without nystagmus but with intermittent esotropia and intermittent head shaking. The head movements manifested only when the eyes were straight and ceased with the spontaneous onset of esotropia or during occlusion of either eye. When the child’s head was forcibly stabilized, he immediately developed esotropia. The authors concluded that the head shaking somehow facili-
tated ocular alignment. A similar case was recently reported by Khan.³

To our knowledge, there has been no further study of this phenomenon, although it is well recognized that head oscillations are used to stabilize the eyes in space and thereby improve vision during periods of visual fixation in patients with spasmus nutans.⁴ Although the fine torsional nystagmus and head oscillations in our patients superficially resembled spasmus nutans, none of our patients displayed the asymmetrical nystagmus or torticollis of spasmus nutans.⁴ Interestingly, a long-term follow-up of children with spasmus nutans showed a 50% incidence of esotropia, latent nystagmus, and dissociated vertical divergence,⁵ suggesting nosologic overlap with the syndrome in our patients.

While nystagmus appears to cause the head oscillations in spasmus nutans, it seems more likely that infantile esotropia was the driving force in our patients, and that the fine torsional nystagmus was simply an epiphenomenon of the infantile esotropia. In our experience, it is not uncommon for slitlamp examination to disclose a subtle torsional nystagmus in infants with uncorrected infantile esotropia who later develop a horizontal latent nystagmus following surgical realignment. This mechanism is supported by the two previous reports of head shaking with intermittent esotropia and no nystagmus.²,³

All 3 of our patients eventually developed a horizontal latent nystagmus with other ocular motor signs of infantile esotropia. In 2 patients (patients 2 and 3), the degree of esotropia was highly variable, but the amplitude of the nystagmus did not vary inversely with the size of the esotropia (ruling out a nystagmus blockage mechanism). In patient 2, the parents noted that they could stop the head oscillations by removing the child’s hyperopic glasses. This observation could indicate that either the head oscillations served to improve a tenuous binocular alignment (if the esotropia was causing the head oscillations) or increased convergence dampened the nystagmus and thereby diminished the head oscillations (if the nystagmus was causing the head oscillations). The degree of associated neurological disease was also variable; 1 child was neurologically normal, 1 had mild global neurodevelopmental delay, and 1 had syndromic hydrocephalus. The trajectory of head oscillations often varied in the same patient, from head shaking to head nodding to elliptical head movements. Surgical realignment of the eyes produced immediate improvement of the head movements in all patients, with complete immediate resolution in 2 and marked improvement followed by rapid resolution in the third.

We conclude that infantile esotropia, when accompanied by nystagmus, can rarely be associated with prominent head oscillations. In this syndrome, surgical realignment of the eyes can extinguish the associated head oscillations. It therefore seems that infantile esotropia, nystagmus, or both must play a causal role in the head shaking or head nodding of neurologically predisposed children. The central mechanism by which infantile esotropia with nystagmus leads to oscillatory head movements is still unknown.

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