Author: Amy Aldrich, BS, OD

Residency: Pediatric Optometry and Vision Therapy; Southern California College of Optometry at Marshall B. Ketchum University

Title: Infantile Esotropia Syndrome with Fusion Maldevelopment Nystagmus Syndrome

Abstract: This case describes an infant with esotropia, both eyes apparently fixed in adduction, and latent nystagmus. Infantile esotropia syndrome and fusion maldevelopment nystagmus syndrome, as well as important differential diagnoses are reviewed.

I. Case History

- 4-month-old Hispanic male
- Chief complaint: bilateral inward eye turn. At least one eye has turned in since birth. Patching 1 hour per day, alternating the patched eye each day.
- Born at 36 weeks gestational age. In the NICU for 1 month following birth. Normal developmental milestones.
- No medications.

II. Findings

- VA: unable to measure, occasional ability to monocularly fixate
- Hirschberg/Kappa: constant bilateral esotropia of approximately 33pd OD/OS
- Versions/ductions: OD and OS in adduction; no abduction, elevation or depression present OD/OS. No palpebral fissure narrowing.
- Latent nystagmus OD/OS
- Dry retinoscopy: fluctuating low refractive error. No impact on eye position with moderate or high plus lenses OU.
- Anterior segment and pupils: normal

III. Differential Diagnosis

- Primary: Infantile esotropia syndrome, fusion maldevelopment nystagmus syndrome
- Others: Ciancia Syndrome, nystagmus blockage syndrome, early-onset accommodative esotropia, bilateral cranial sixth nerve palsy, strabismus fixus

IV. Discussion
• Infantile esotropia syndrome describes a large-angle esotropia with onset in the first 6 months that is unresponsive to plus lenses in a neurologically normal child. Common associated findings are oblique muscle dysfunctions, DVD, AV syndromes, nystagmus, and cross-fixation.

• Fusion maldevelopment nystagmus syndrome (FMNS) describes a latent nystagmus of infantile onset that is associated with strabismus. The nystagmus may change with exaggerated convergence (“blockage”), and an anomalous head turn is often associated with the fixing eye in adduction.

• Ciancia Syndrome is a condition that can accompany a large-angle infantile esotropia. Patients manifest a bilateral limitation of abduction, a jerk nystagmus with attempts to abduct, and a face turn or head tilt towards the fixating eye.

• Nystagmus blockage syndrome occurs when an infantile nystagmus and infantile esotropia are concomitant. A willful esotropia and convergence-like eye movements are made to dampen the nystagmus.

• To rule out a bilateral sixth nerve palsy when no abduction is observed on initial testing, prolonged patching or the doll’s head maneuver can confirm the ability to abduct. If the direction of the nystagmus switches based on which eye is fixing, then the nystagmus is due to FMNS rather than a neurologic condition.

V. Management

• Prompt referral to a pediatric ophthalmologist, who diagnosed concurrent Ciancia Syndrome and nystagmus blockage syndrome. Further review of the case supports a more likely diagnosis of infantile esotropia syndrome with FMNS.

• Patching 2 hours per day, alternating the patched eye each day

• Per consulting ophthalmologist, surgical correction of esotropia will be at 1 year of age when infant can better tolerate anesthesia.

• Current research indicates the need for much earlier correction of the esotropia to improve surgical outcome. Typically, monofixation syndrome is the best expected outcome.

VI. Conclusion

Infantile esotropia syndrome and FMNS are important conditions that present in infants. The differential diagnoses are numerous, and the practitioner must rule-out serious neurological conditions. Management includes occlusion therapy to prevent amblyopia and prompt co-management with a pediatric ophthalmologist for appropriate surgical treatment.