The Impact of Gaze Deficits in Children with Cerebral Palsy

Lawrence Tychsen, MD
Washington University in St. Louis
St. Louis, MO

LEARNING OBJECTIVES
1. The attendee will be able to describe common gaze deficits in children with CP.
2. The attendee will be able to explain the relationship between the prevalence of gaze deficits and level of severity of CP.
3. The attendee will be able to identify tests that distinguish gaze deficits from cerebral visual impairment.

CME QUESTIONS
1. What are the most common gaze deficits observed in children with CP?
2. What is the Gross Motor Function Classification Scale and why is it of value to the neuro-ophthalmologist?
3. What is the definition of CP?

KEYWORDS
1. Gross Motor Function Classification Scale
2. gaze apraxia/palsy
3. fixation impersistence
4. cerebral visual impairment
5. cerebral palsy

INTRODUCTION
Cerebral palsy (CP) is a permanent, nonprogressive disorder of movement and posture due to a lesion of the fetal or infant brain. The goal was to determine whether children with different severities of CP, as defined using the Gross Motor Function Classification System (GMFCS), had different degrees or types of visual dysfunction.

METHODS: An observational, cross-sectional-design study was conducted by using neurologic and masked ophthalmic measurements on a representative cohort of 50 children with CP.

Mean age was 5.6 years (range, 2-19.5 years); mean gestational age was 31 weeks.

RESULTS: The likelihood of debilitating visual deficits was greater in children with higher GMFCS scores, independent of gestational age. Children with level 5 disease (most severe) were at greatest risk for high myopia, absence of binocular fusion, dyskinetic strabismus, severe gaze dysfunction, and optic neuropathy or cerebral visual impairment (CVI). These deficits were rare or absent in children with the mildest disease, level 1. Cataloging of gaze dysfunction included the following descriptors.

Gaze apraxia/palsy: difficulty initiating saccades in response to a step change of target position, evident as abnormally long latencies and/or subnormal saccade amplitude; apraxia connotes disfacility more severe for performance on command, and less severe for spontaneous saccades, including persistent gaze deviations away from primary position in the orbits; supranuclear origin of the palsy is verified by demonstrating full eye rotations on vestibulo-ocular reflex testing (doll’s eyes).

Pendular-jerk nystagmus: oscillation of eye position under conditions of binocular viewing while fixating a stationary target, with pendular or increasing-velocity slow phases, depending on gaze position in the orbits; each cycle of increasing velocity slow phase jerk nystagmus is followed by an oppositely directed fast-phase saccade.

Fixation impersistence: noncyclical instabilities of eye position in subjects attempting to view a stationary target; manifested as saccades 5 deg or larger directing the eyes away from target position, and/or slow-phase drifts, not followed by refoveating saccades.

Gaze disorders that are features of infantile-onset strabismus—pursuit/OKN asymmetry and latent nystagmus—were common, evident in at least 20% of the children in all GMFCS levels, with the children in levels 2 to 5 showing the highest prevalence. Seventy percent of the children in level 1 had no gaze disorder. The highest rates of more severe gaze dysfunction i.e., pendular-jerk nystagmus, gaze apraxia/palsy, and fixation impersistence—were detected in the children of CP level 5, affecting 30% to 40% (P = 0.03). Likewise, children with quadriplegic disease had more dyskinetic strabismus (P = 0.04) and more severe gaze dysfunction in the form of fixation impersistence (P = 0.04).

CONCLUSIONS: Children with higher level GMFCS scores of CP have greater gaze dysfunction. Gaze dysfunction should be distinguished from the visual sensory deficits that typify CP.
DISCUSSION
Observations of gaze dysfunction in CP have been described using a variety of terms: uncoordinated saccades and pursuit; paroxysmal ocular deviations; stability of fixation inability; struggling with fixation and eye movement; complete disruption of ocular motor organization; dyskinetic eye movement disorder; and ocular motor apraxia. Jan et al. (2001) reported that one or more of these deficits was present in all 14 children chosen for study because of severe, dyskinetic CP. Salati et al. (2002) stated a prevalence of 78% in children categorized as having spastic or mixed, moderate-to-severe quadriplegia.

The results of our study indicate lower prevalences of gaze dysfunction: an average 22% in children with quadriplegic CP; an average 18% in children of CP levels 3 to 5; and an average 8% for levels 1 to 2. Irrespective of discrepancies in prevalence, conjugate gaze deficits deserve better recognition as significant components of visual impairment in children with CP, as they often masquerade as CVI.

We were struck by the number of children in our cohort who had normal or mildly subnormal visual acuity, but who, on review of outside medical records, had been labeled as having “cortical blindness.” Parents, therapists, and other healthcare professionals described them variously as: “visually inattentive,” “unable to see or follow,” “no eye contact,” “looks away,” “looks over,” or “uses only peripheral vision.” Careful clinical observation and eye movement recordings revealed combined saccadic and smooth pursuit gaze apraxia/palsy, and fixation impersistence. The children with these deficits who retained head and neck control used horizontal or vertical head thrusts to facilitate gaze shifts. Restraining head motion tended to exacerbate the impairments by prolonging latencies, reducing saccade amplitudes, and reducing pursuit/OKN velocity matching. The deficits impaired both horizontal and vertical gaze, but were often more pronounced for vertical target steps and target motion.

CME ANSWERS
1. The most common gaze deficits in children with CP are: gaze apraxia/palsy, pendular–jerk nystagmus, and fixation impersistence.

2. The Gross Motor Function Classification System (GMFCS) is a standardized scale for grading the severity of motoric impairment in CP. It ranges from Level 1: mild impairment of walking, to Level 5, severe limitation requiring a transporter device.

3. CP is a permanent, nonprogressive disorder of movement and posture due to a lesion of the fetal or infant brain.

REFERENCES
