Chronic Progressive External Ophthalmoplegia and Bilateral Vestibular Hypofunction: Balance, Gait, and Eye Movement Before and After Multimodal Chiropractic Care: A Case Study

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ABSTRACT

Objective: The purpose of this report is to describe care of a patient with chronic progressive external ophthalmoplegia and bilateral vestibular hypofunction.

Clinical Features: A 66-year-old patient presented with limited eye movement and mild ptosis, which led to a diagnosis of chronic progressive external ophthalmoplegia. Rotary chair testing suggested vestibular involvement. Other symptoms included dizziness, problems with balance, and chronic stiffness in his cervical and thoracic regions. He had anxiety about loss of function and limited exercise habits because of fear of falling. Examination methods included balance assessment, kinetic aspects of walking, and videonystagmography.

Intervention and Outcome: He had already begun regular practice of vestibular rehabilitation exercises before receiving 18 sessions of manual and instrument-assisted chiropractic manipulation, along with mobilization, stretching, and transverse massage, over 37 weeks. In addition to self-reported improvements, there was substantially decreased postural sway during balance assessment and there were small improvements in eye movement, ptosis, and walking.

Conclusion: This patient showed improvements in balance, eye movements, and walking while undergoing multimodal chiropractic care and practicing eye and balance exercises. (J Chiropr Med 2019;18:144-154)

Key Indexing Terms: *Bilateral Vestibulopathy; Eye Movement Measurements; Postural Balance; Manipulation, Chiropractic; Ophthalmoplegia, Chronic Progressive External*

INTRODUCTION

Chronic progressive external ophthalmoplegia (CPEO) is a form of mitochondrial disease characterized by ocular motor deficit and progressive ptosis, both often symmetrical; ptosis is usually the first sign.¹⁻³ Often patients experience progressive paralysis of the levator palpebrae, orbicularis oculi, and extraocular muscles, especially for upward gaze.^{4,5} As ptosis worsens, the patient may attempt to use muscles of the face to try to lift the eyelids or may tilt the head backward to see; as eye muscles become weaker,

the patient may have to turn the head to see in different directions.⁶ Weakness of arm, leg, and neck muscles may accompany disease progression.⁶ Exercise intolerance may result from myopathy in the arms or legs, and dysphagia may result from myopathy in the neck.⁶ Some patients experience additional signs and symptoms, including sensorineural hearing loss or ataxia, and may be classified as having “progressive external ophthalmoplegia plus.”⁶

Although not common, CPEO should be considered when patients present with ocular myopathy. In most cases, definitive diagnosis is by muscle biopsy, positive for atrophy and ragged-red fibers (muscle cells containing excess mitochondria).⁴⁻⁷ However, magnetic resonance imaging is increasingly being used to confirm muscle atrophy.⁸

In a review, Fraser³ found no evidence supporting any treatment as being effective for CPEO. In general, there is little beyond anecdotal evidence supporting any treatments for the management of mitochondrial diseases.^{1,9-12} No medications have been approved by the US Food and Drug Administration,¹² and there is little evidence that nutritional

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supplements are effective⁹; medications and supplements will be discussed further later. Options for addressing secondary issues are limited^{1,2,4} for CPEO; the most common is corrective surgery for ptosis, although a nonsurgical alternative is to have ptosis crutches affixed to a patient's glasses.³ In general, patients with vision impairment may fear further loss and subsequent reductions in their independence. Their emotional well-being may diminish because of reduced social functioning.¹³

Clinicians should recognize that other syndromes may be present to identify factors that influence quality of life and to provide symptomatic treatment where possible. Awareness of atypical presentations in CPEO may help. Ramakrishnan⁷ reported a CPEO case with vocal cord palsy. Bucelli¹⁴ reported on 2 cases without ptosis. Caballero⁴ reported 6 cases. Half of the cases had asymmetric ptosis, half had mild dysphagia, 2 also experienced migraine headaches, and all but 1 had ragged-red fibers on biopsy.⁴ Smits¹⁵ reported 28 CPEO cases, many with severe fatigue, depression, pain, and other functional impairments.

Vestibular hypofunction is characterized by a reduced or absent functioning of the vestibular organs or the vestibular nerves or both. The problem may be unilateral vestibular hypofunction or bilateral vestibular hypofunction (BVH).¹⁶ It is often idiopathic but can be related to genetic disorders, Ménière's disease, infectious diseases, neurodegenerative diseases, or ototoxicity.¹⁶ Ototoxicity can be an adverse effect of aminoglycoside antibiotics, including gentamicin, tobramycin, amikacin, streptomycin, neomycin, kanamycin, paromomycin, netilmicin, and spectinomycin.¹⁷ Individuals affected by BVH may experience postural instability, visual blurring during head movement, or dizziness, and their fall risk is greater.^{18,19} Many patients with BVH develop depression, anxiety, or both.^{18,19} A retrospective study of BVH patients noted that 36% also had "joint problems" and 22% complained of neck pain.¹⁸

Agrawal²⁰ determined that 35.4% of adults 40 years and older in the United States have some degree of vestibular dysfunction and that the incidence increases with age. Ward²¹ calculated that loss or reduction of vestibular function is bilateral for approximately 28 of every 100 000 people in the United States. Many, but not all, patients with vestibular dysfunction will improve with vestibular rehabilitation, particularly exercises designed to improve postural stability during head movements.^{19,22}

The purpose of this report is to describe the chiropractic management of a patient who had previously been diagnosed with CPEO complicated by "probable" BVH.

CASE REPORT

Clinical Features

A 66-year-old male patient presented with the chief complaints of dizziness, light-headedness, and balance

problems. Previously, in 2011, an ophthalmologist noticed a smaller range of eye movements than seemed typical. A brain magnetic resonance imaging turned up nothing abnormal and he had no evident impairment, so no further investigation was done at that time. In 2015, his internist recommended Zyrtec and Flonase for some occasional light-headedness, which cleared up quickly—however, it returned within a few weeks. At some point he began to notice a small amount of hand tremor, enough to affect his handwriting, and occasional mild head tremor. Subsequently, he was examined by 2 otolaryngologists, an otologist, and 2 neurologists; they ruled out allergies and vertigo as causes of the light-headedness and ruled out Parkinson disease regarding the tremors. He also underwent cataract surgery in both eyes that same year.

Later, a neurotologist saw the limited range of eye movement and diagnosed him as having CPEO. Although a definitive diagnosis of CPEO would involve biopsy of the eye muscles,^{4,7} the neurotologist was so certain of his diagnosis he recommended against the biopsy. He did note, however, that rotary chair testing suggested vestibular system involvement and offered BVH as a probable additional diagnosis. The patient was referred for balance testing and treatment by a physical therapist, who taught him several exercises, such as heel-walking and toe-walking; walking in a straight line while rapidly looking left and right; and sitting with his head centered and immobile while looking left, right, up, and down. Additional recommendations included activities to challenge his balance, such walking on uneven surfaces, hiking, and racket sports. No diagnosis was made for the tremors, and he has had no treatment for them.

Understanding that there was no established treatment for CPEO and fearing further loss of function, the patient sought out chiropractic care in an effort to make any improvement possible. At the time of his presentation to our research center, features of CPEO were present only in a mild form and were not obvious. He had a slightly droopy appearance of his eyelids and a mild furrowing of the brow, related to mild ptosis and use of facial muscles. His light-headedness had improved somewhat, presumably attributable to the exercises he had learned from the therapist; however, he was having noticeable problems with balance and dizziness. His exercise habits, mostly walking and golf, had been limited because of fear of aggravation of symptoms. In his administrative job, he needed to sort through hundreds of emails each day. This aggravated his light-headedness, and he concluded that he was using repetitive head movements to compensate for limited eye movement. He had begun taking Zolofit (sertraline hydrochloride) (Pfizer Inc, New York, New York) approximately 2 weeks earlier because of anxiety and fear about his condition.

He also had chronic stiffness in his lower cervical and upper thoracic region and low back, right "shoulder" pain (lower neck, upper thorax, and anatomical shoulder), and

Fig 1. BalanceCheck Mobile Screener portable balance screening system. (A) The force plate itself can be seen in a normal stability test. (B) The 3-inch-thick cushion used to create a “perturbed” condition as a challenge to stability.

some sense of leg weakness. The treating doctor of chiropractic accepted his case on the premise that chiropractic care might offer some benefit, at least for his pain and stiffness, and possibly for some other symptoms. Before examination and treatment began, we obtained approval from the research ethics board of Life University.

Outcome Assessments

Biomechanical and neurologic evaluations included assessments of postural sway with a portable force plate, eye movements with a computerized binocular goggle system, and gait evaluation with an instrumented treadmill. These were done before care; again after 12 chiropractic visits (week 20); and in a final session, after 6 more visits (week 37).

Postural sway during static standing was evaluated with a BalanceCheck Mobile Screener portable balance screen-

Fig 2. VisualEyes binocular goggles used for videonystagmography.

ing system (Bertec Corp, Columbus, Ohio). In a “normal” stability test the patient stood still with his feet apart (Fig 1A), first with eyes open and then with eyes closed. Then, he stood on a 3-inch-thick cushion (Fig 1B) in a “perturbed” condition, which presents a challenge to stability, with the eyes open and closed, and with the head turned left, turned right, flexed forward, and extended backward. The system software measures range and direction of sway to calculate a stability score.

Video nystagmography was performed with VisualEyes binocular goggles (Fig 2; Micromedical Technologies, Chatham, Illinois). During an exam, patients watch dots of light move in various patterns through their visual fields while instrumentation in the goggles record eye movement. We evaluated accuracy, latency, and velocity of saccadic movements; gain of smooth pursuit movements; and optokinetic response.

For visual perception to be at its clearest, images need to be projected on the fovea centralis of the macula²³; that goal is accomplished differently for stationary and moving objects. Saccades are ballistic movements in which the eyes move from one stationary location to another, such as in reading or looking around a room. Once the brain determines where a visual target is and how far the eye needs to move, “latency” is a measure of the time it takes. Saccades can also be measured in amplitude, velocity, and duration, which help to determine accuracy of target location. Accuracy may be affected by dysmetria, either in the form of overshoot (too far) or undershoot (not far enough). Large and frequent overshoot is abnormal but rare; undershoot is common in normal participants.²³⁻²⁵

In smooth pursuit, eye movement accelerates to catch up to a moving target and then decelerates to match its

Table 1. Stability Scores for the Bertec BalanceCheck Mobile Screener

	NS-EO	NS-EC	PS-EO	PS-EC
Age-matched norm SS	91.7	90.0	85.4	75.2
Pre-care SS	88.1	88.7	75.5	52.4
Midcare SS	92.6	92.5	86.4	82.5
Postcare SS	95.7	91.5	90.6	81.2

Age-matched norms are averages provided by the proprietary software for clinical comparison.

EC, eyes closed; EO, eyes open; NS, normal stance; PS, perturbed stance; SS, stability score.

velocity. The ratio of peak eye movement velocity to target velocity is calculated as *gain*. Latency is a measure of the time required. Smooth pursuits work well for slower-moving targets but have trouble tracking very fast movements. The limitation can be compensated for in 2 ways. If a movement pattern is predictable, a smooth pursuit can be initiated early; for movements that are unpredictable, smooth pursuits are supplemented with saccades.²³⁻²⁵ What is known as the optokinetic (OPK) response involves a combination of saccades and smooth pursuits. For example, in a situation of continual movement in one direction, the eyes smoothly follow from one point to another, flick back to the starting position,²⁶ and then repeat the process.

Gait Analysis. A treadmill with force sensors mounted underneath the walking belt (Noraxon, Scottsdale, Arizona) was used to record spatial and temporal gait parameters and vertical ground reaction forces (VGRFs). The patient walked at various speeds for a few minutes to determine his preferred walking speed, a speed at which variability is likely at its lowest.^{27,28} He was then asked to walk “as consistently and symmetrically as possible” for 2 trials, each 1 minute, and rested in between.

The system’s standard clinical report provides parameters such as step length and width, and proportions of gait cycle stance and swing phases. Vertical ground reaction forces were evaluated with a custom program written by the second author (R.S.H.) in Visual Basic (Microsoft, Redmond, Washington). Variability of specific events—peak forces in loading, midstance, and push-off, and the timing of those events—was calculated by coefficients of variation.

Examination and Chiropractic Intervention. An examination appropriate for spine and upper-extremity mechanical dysfunction was done: inspection of posture, spinal range of motion, palpation, manual muscle testing, and provocative tests. Briefly, the findings suggested articular dysfunction at multiple cervical and thoracic spinal levels, multiple areas of muscle hypertonicity, grade 4 weakness of the right deltoid, and suspected right supraspinatus tendinosis. Chiropractic manipulation (spinal adjustments)

consisted of a combination of mobilization; drop-table assisted high-velocity, low-amplitude thrusts; and instrument-assisted adjustments by use of an Impulse instrument (Neuromechanical Innovations, Chandler, Arizona) directed toward joints determined to lack normal intersegmental mobility and alignment. Choices of methods were a combination of practitioner and patient preferences. Most spinal manipulative thrusts were directed toward the C1-3, C7-T1, and T5-7 areas, all judged to have chronically restricted and aberrant motion. Some visits included mobilization or adjustment of the scapula, the glenohumeral joint, one or both innominates, the sacrum, and other spinal segments.

Most visits included short-duration stretching of the upper trapezius, levator scapula, and other muscles of the cervical region and transverse massage in some muscles with palpable trigger points or fibrotic areas, including the upper and middle trapezii, levator scapula, supraspinatus, infraspinatus, teres major and minor, subscapularis, pectoralis major and minor, coracobrachialis, latissimus dorsi, deltoid group, and both heads of the biceps—bilaterally, for most, but with more focus on the right side. The treating doctor of chiropractic also added one therapeutic exercise to the patient’s existing regimen: shoulder circumduction performed bilaterally, with both sides moving simultaneously in opposite directions, while looking straight ahead.

Eighteen sessions of care were done in 37 weeks. The first 7 visits were done 1 to 2 times per week followed by a 6-week gap related to holidays and vacations; then 5 visits were done at 1-week intervals, with the remaining 6 visits scheduled approximately 2 weeks apart.

Patient-Reported Responses to Care

After the first few visits, the patient had no additional problems with light-headedness, dizziness, or balance. He resumed regular hiking without fear. Shortly before his final visit, he reported hiking on a moderately challenging trail with no difficulty. He was able to continue playing golf. He reported no falls during the 37 weeks of active care, at a follow-up visit 18 months later, and during a follow-up phone call 2½ years after the first assessment. At the 18-month follow-up, he was mostly concerned with neck and shoulder tightness. He no longer has problems with reading email, but also went through a scheduled retirement and no longer has the same responsibilities.

Postural Sway

Stability scores for the pre-care, midpoint, and postcare assessments and age-matched average stability score, may be viewed in Table 1. Higher scores indicate better stability. Pre-care, the patient was below his age-matched averages by more than 3 standard deviations for all conditions: normal stance-eyes open, normal stance-eyes

closed, perturbed stance-eyes open (PS-EO), and perturbed stance-eyes closed. By midpoint, he had improved to exceed all averages. Postcare, he improved further in normal stance-eyes open and PS-EO but showed small dips in normal stance-eyes closed and perturbed stance-eyes closed. The PS-EO graphs in Figure 3 are scans of printouts created by the BalanceCheck system. Larger ovals describe greater sway patterns. The proprietary software has no option for graph customization; for meaningful visual comparison, we cropped and scaled the images to equivalent sizes.

Video Nystagmography

The results of video nystagmography were mixed. Some features showed improvement from pre-care to postcare; other features were unchanged or worsened. Overall, the results, both pre- and postcare, show better visual tracking in vertical directions, as compared with horizontal.

During the horizontal and vertical saccade exams, a dot of light randomly appeared at various points across the field of vision. Pre-care, for horizontal saccade accuracy, both the left and right eyes showed a tendency to overshoot a target when moving to the right: 194% left eye, 105% right eye (perfect accuracy would be 100%).²⁵ Otherwise, both eyes displayed undershoot. Overall, horizontal saccade accuracy was decreased (worse) from pre- to postcare for both eyes. (Table 2). For latency, pre- to postcare showed improved tracking (shorter times) to the left in both eyes and tracking to the right in the right eye; there was a decline (increased time) in the left eye tracking to the right. Horizontal saccade velocity, in degrees per second, showed a decrease (worsened) in both eyes from pre- to postcare. However, postcare accuracy, latency, and velocity all showed more symmetry between the left and right eyes compared with pre-care. Vertical saccade accuracy displayed undershoot for both pre- and postcare. Postcare, there was an increase in accuracy for the left eye tracking upward and the right eye tracking downward. Conversely, there was decreased accuracy for the left eye tracking downward and the right eye tracking upward. Latency in both eyes was decreased (improved) when tracking upward and increased when tracking downward. Vertical saccade velocity was slower (worsened) from pre- to postcare in both eyes. Pre-care, vertical saccade accuracy, latency, and velocity were all asymmetrical for both eyes; latency and velocity continued to show asymmetry postcare.

Fig 3. Graphs for the PS-EO condition, in which the proprietary software provides smoothed depictions of CoP anterior-posterior and lateral sway (scanned from printed reports). (A) Pre-care, (B) midpoint, and (C) postcare. Variances in visual appearance reflect how the system automatically formatted differently from the first graph to those later.

Table 2. Saccades

		Pre-care		Midcare		Postcare	
Horizontal saccades *		Left eye	Right eye	Left eye	Right eye	Left eye	Right eye
Accuracy (%)	Leftward	68	85	62	66	54	50
	Rightward	194	105	55	83	50	45
Latency (ms)	Leftward	378	416	342	391	371	375
	Rightward	290	368	361	386	361	353
Velocity (°/sec)	Leftward	301	124	130	123	43	32
	Rightward	359	153	122	138	36	28
Vertical saccades		Left eye	Right eye	Left eye	Right eye	Left eye	Right eye
Accuracy (%)	Upward	52	59	67	58	55	53
	Downward	93	72	113	91	87	78
Latency (ms)	Upward	295	307	256	265	258	283
	Downward	288	263	242	330	310	327
Velocity (°/sec)	Upward	81	78	104	107	70	54
	Downward	217	156	260	195	122	111

Accuracy is the precision in which the eyes locate a target. Ideal accuracy is 100; higher percentages represent overshoot, and lower percentages represent undershoot. Latency is the time between the target movement and the beginning of a saccade. Velocity is the speed at which the eye moves to the target (degrees/s)

* Normative values for horizontal saccades, from healthy volunteers in the range of 61 to 70 years old, were reported by Seferlis: accuracy, 88-89; latency, 290-293; and velocity, 170-190.²⁹

For the smooth pursuit exams, a dot of light smoothly moved across the field of vision at motion frequencies of 0.1 Hz, 0.2 Hz, and 0.4 Hz (Table 3). Data were recorded for horizontal and vertical gain. Horizontal gain was low

pre-care for both the left and right eye and showed minimal change at the final assessment. Vertical gain, however, began with values closer to 1.0 (ideal) than horizontals and had postcare increases (improvement) at all 3 frequencies.

Table 3. Smooth Pursuits

		Pre-care		Midcare		Postcare	
Gain: horizontal *		Left eye	Right eye	Left eye	Right eye	Left eye	Right eye
0.1 Hz		0.43	0.45	0.38	0.38	0.41	0.40
0.2 Hz		0.37	0.41	0.36	0.45	0.36	0.39
0.4 Hz		0.28	0.28	0.28	0.37	0.30	0.29
Gain: vertical		Left eye	Right eye	Left eye	Right eye	Left eye	Right eye
0.1 Hz		0.78	0.77	0.88	0.98	0.89	0.92
0.2 Hz		0.80	0.74	0.87	0.78	1.08	1.10
0.4 Hz		1.01	0.88	0.90	0.68	1.01	1.01

Gain represents how closely the target is being tracked. Ideal gain is 1.0.

* Horizontal smooth pursuit gains were reported by Seferlis to be in the range of 0.79 to 0.81 in healthy volunteers aged 61 to 70.²⁹

Table 4. *OPK Exam*

	Pre-care		Postcare	
Gain: horizontal	Left eye	Right eye	Left eye	Right eye
Leftward	0.12	0	0	0
Rightward	0	0	0	0
Gain: vertical	Left eye	Right eye	Left eye	Right eye
Upward	0	0	0.39	0.32
Downward	0.45	0.60 ^a	0.37	0.37

Gain represents how closely the target is being tracked. Ideal gain is 1.0. A gain of zero indicates no movement was detected.

OPK, optokinetic.

^aMeasurement of downward gain of the right eye, pre-care, was affected by a drooping eyelid and eyelashes.

During the OPK exam, columns or rows of dots cascaded across the field of vision such that the eyes move 30° per second. Pre-care, the OPK exam made the patient feel disoriented and nauseous. The exam was not attempted at midcare, but at the postcare session he was able to tolerate it. Pre-care video recordings show very little eye movement, which is reflected in the small and zero gain values in Table 4. For horizontal gain there was little change from pre to post, with a decline for the left eye tracking leftward. For vertical gain, both eyes showed improvement in tracking upward, but there was a decline in left eye tracking downward. The pre-

care downward gain measurement is thought to be faulty because of right eyelid ptosis (Fig 4A), and the system software had difficulty tracking the pupil; the measurement of 0.60 appears to be movement of his eyelashes. Also, as detected from the postcare video, both eyes were more open and alert and had better stamina (Fig 4B).

Gait Analysis

Proportions of stance phase and double stance were relatively high and swing phase relatively low, suggesting suboptimal walking stability; these did not change postcare (Table 5). However, by the time of the patient’s final assessment, his self-selected walking speed, stride length, and cadence had increased, and step width decreased. These small changes, coupled with his subjective experience, suggest improved stability. Pre-care VGRFs (not shown) showed moderately high variability in the timing of peak forces and mid-stance. Postcare, the VGRF results were mixed and showed no clear trend toward improvement or worsening.

The patient provided consent for this case report to be published.

DISCUSSION

There currently is no established treatment for CPEO, a form of mitochondrial disease, and there have been no previous reports of chiropractic care for either CPEO or

Fig 4. (A) A typical appearance of the eyes during the pre-care optokinetic evaluation; note the drooping of the right eyelid. The system software identified the pupil with a + symbol but had difficulty with the right pupil being obscured by eyelashes. (B) The eyes during the postcare OPK evaluation. OPK, optokinetic.

Table 5. *Spatiotemporal Parameters of Walking Gait*

Parameter	Pre-care	Midcare	Postcare
Stance phase, left, %	67.4 (1.5)	68.2 (1.6)	67.7 (1.2)
Stance phase, right, %	66.9 (1.7)	67.5 (1.8)	66.9 (1.7)
Swing phase, left, %	32.6 (1.5)	31.8 (1.6)	32.3 (1.2)
Swing phase, right, %	33.1 (1.7)	32.5 (1.8)	33.1 (1.7)
Double support, %	34.3 (1.6)	35.7 (1.6)	34.6 (1.5)
Single support, left, %	33.1 (1.7)	32.5 (1.7)	33.1 (1.7)
Single support, right, %	32.6 (1.6)	31.8 (1.6)	32.3 (1.4)
Stride length, cm	96.2 (2.5)	94.1 (2.5)	102.7 (2.0)
Step width, cm	16.7 (2.0)	17.5 (2.0)	15.1 (2.0)
Cadence, step/min	99.8 (2.0)	97.5 (2.0)	102.7 (2.0)
Velocity, km/h	2.9 (0.1)	2.8 (0.1)	3.2 (0.1)

Data presented as means (standard deviations).

BVH. This patient showed substantial improvement in balance while undergoing chiropractic care and practicing eye and balance exercises. He also had small improvements in eye movements, right eye ptosis, and walking. Although his diligent practice of eye and balance exercises may be the most important component, the improvements came after beginning multimodal chiropractic care. He had less anxiety and fear about loss of independence—partly because of his improved function and partly, as he stated it, due to coming to grips with his own finality. To the degree that either chiropractic care or the exercises had an impact, it could more likely be with BVH given the pathophysiology of CPEO. Also important is that his CPEO was not severe and no progression has been detected.

Our gait protocol encourages selection of a walking speed that can be safely maintained; people with instability may naturally choose slower speeds^{30,31} and wider steps.^{31,32} Not surprisingly, walking speed may be slower with vestibular deficiencies.³³ The postcare self-selected faster speeds and narrower step widths seen in this report may seem like small changes, but step width decreased by 9.6% (1.6 cm), and his 1.1% increase in walking speed was similar to the 1.2% reported by Herdman¹⁸ for patients with BVH.

Although the video nystagmography postcare results were mixed, there was some overall improvement. The patient was able to tolerate the OPK exam at the final assessment with less eye fatigue. And he had visible improvement in ptosis of the right eyelid.

The findings of other studies may provide perspective. The CPEO cases without ptosis reported by Bucelli¹⁴ had “markedly slow” saccades. DeBrosse³⁴ described a truck driver with CPEO and severe ptosis who used adhesive tape

to retract his eyelids. That patient’s horizontal and vertical saccades were slow, and he lacked smooth pursuit movements—his eyes used multiple small saccades to track moving targets.³⁴ Shinmei³⁵ reported on 3 patients with mitochondrial encephalopathy with lactic acid and stroke-like episodes. All 3 patients had horizontal and vertical saccadic undershoot, with prolonged latency, and low gain of smooth pursuits, with frequent “catch-up” saccades.³⁵ Fahnehjelm³⁶ reported on 13 patients with mitochondrial complex I deficiency, and many of them had “saccade dysfunction” and asymmetry of smooth pursuits.³⁶

At present, no effective treatments have been established for mitochondrial diseases.¹⁻⁴ There is hope for gene therapy.³ Some medications are used off-label,³⁷ and there are some investigational medications, such as KL1333,³⁸ bezafibrate, resveratrol, dimethylglycine, and EPI-743, a synthetic analog of vitamin E.¹² Several sources suggest coenzyme Q₁₀ may help,^{3,11,12,39-43} and a synthetic analog of coenzyme Q₁₀, idebenone, has been investigated for Leber’s hereditary optic neuropathy, another mitochondrial disease that affects the eyes.¹ Otherwise, there is little evidence,^{3,42} so far, for a number of nutritional supplements that have been investigated, including thiamine, vitamin E, vitamin K, cysteine, and succinate,¹² riboflavin,^{12,41-43} vitamin C,^{3,12,41-43} vitamin D,^{41,42} arginine,^{12,43} carnitine,^{12,41} creatinine,^{12,39} lipoic acid,^{12,39} melatonin,⁴⁴ mushroom derivatives,⁴⁵ or “cocktails” of multiple supplemental nutrients.^{42,46} There is some support for carbohydrate-restrictive diets.^{42,47-49}

The review by Fraser et al³ stated, “Symptomatic treatments should be considered in all patients with mitochondrial disease to prevent secondary morbidity and

to improve quality of life.”³ Those authors also suggested low vision aids to help compensate for severe vision loss, aerobic exercise training to help with muscle dysfunction and general quality of life, diet modification and insulin for patients who also have diabetes, seizure medication for patients with epilepsy, pacemakers for patients with cardiac conduction defects, cochlear implants for patients with sensorineural hearing loss, and antidepressant and antipsychotic medications for related psychiatric symptoms.³ Similarly, but more simply, Smits¹⁵ recommended a multidimensional approach aimed at specific needs of individual patients, which may include rehabilitation therapy and medication.

In a similar vein, we suggest chiropractic care should be considered a form of tertiary prevention, that is, treatment aimed at maintaining the best functioning as possible in patients who cannot become free of disease or symptoms.⁵⁰ In this context, we consider care provided by chiropractors to not only include manipulation and other forms of manual therapy, but also advice regarding exercise, lifestyle, diet, nutritional supplements, body mechanics and ergonomics, and stress reduction.⁵¹⁻⁵⁵ At present, there appears to have been nothing published regarding chiropractic care with CPEO or other mitochondrial diseases.

There have been no previous reports for chiropractic adjustment or manipulation and BVH. However, there have been reports concerning adjustment or spinal manipulation in cases that may also involve the vestibular system and have some similar symptoms, such as cervicogenic dizziness,⁵⁶ vertigo,⁵⁷⁻⁶⁰ Ménière’s disease,⁶¹ and motion sickness.⁶² If manipulation can have an impact on BVH, the mechanism may not be well understood or established. In a review addressing manual therapy, vestibular rehabilitation, and cervicogenic dizziness, Lystad⁶³ suggested that abnormal afferent input from mechanoreceptors in the cervical spine could create a sensory input disparity between the cervical spine and vestibular system. The disparity could result in symptoms such as dizziness, unsteadiness, and visual disturbances, and manual therapy could help correct the mismatch.⁶³ Morningstar⁶⁴ discussed how input from the visual and vestibular systems, and mechanoreceptors of the joints and soft tissue, might interact with the spine and upright posture. Citing Wyke,⁶⁵ they implied a role for manipulation with vestibular dysfunction, saying, “. . . when the cervical facet joints are experimentally immobilized in the presence of vestibular dysfunction, postural instability becomes apparent. . . stability is restored when the facet joints are mobilized.”⁶⁴

Limitations

As with any report involving a single case, no determination of cause and effect can be made. The authors cannot claim that chiropractic care was responsible for the patient’s improvements, although he did feel that he benefitted from the care. Nor

do the authors claim that their methods are superior or more appropriate than what other practitioners would recommend; others may have better insights or patients with different needs.

We failed to use any kind of functional questionnaire to quantify improvement in activities of daily living or quality of life. The patient was questioned on each visit, however, and he subjectively reported improvement. The study would have been better with appropriate questionnaires.

We did not have direct access to the clinic records for the neurologist who diagnosed BVH; instead, we depended on oral reports from an occupational therapist familiar with the case. We might have obtained additional information from the records, but we have no reason to think anything the therapist told us is incorrect. More investigation would be needed to determine the appropriate role of chiropractic care in patients with these diagnoses.

CONCLUSION

This patient showed substantial improvement in balance while undergoing multimodal chiropractic care and practicing eye and balance exercises and had small improvements in ptosis, eye movements, and walking. These modes of care could function as symptomatic or supportive treatment for patients with CPEO and could help resolve vestibular hypofunction.

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CONTRIBUTORSHIP INFORMATION

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Supervision (provided oversight, responsible for organization and implementation, writing of the manuscript): B.S.R., R.S.H.

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Practical Applications

- Chronic progressive external ophthalmoplegia, although uncommon, has no established treatment.
- There have been no previous reports of chiropractic care with these conditions.
- A patient received chiropractic and rehabilitative exercises that helped with quality of life.

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