

Convergence Exercises for Convergence Insufficiency Type Exotropia in Bilateral Internuclear Ophthalmoplegia

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Abstract: *Purpose;* To report a case of successfully treated convergence insufficiency type exotropia (CI-XT) in the context of bilateral internuclear ophthalmoplegia (BINO) using only convergence exercises.

Observations; A 17-year-old male with a history of a resected posterior fossa pilocytic astrocytoma presented with constant diplopia since the craniotomy. Patient and his parents were adamant in pursuing surgical treatment for his new onset strabismus. With appropriate counseling and management with convergence exercises only, the CI-XT was successfully treated without surgery over the course of two years.

Conclusion and Importance; We present a non-invasive approach to the treatment of CI-XT in the context of a BINO, also known as wall-eyed BINO (WEBINO). This approach should be considered before any surgical intervention, especially if prism is unable to successfully neutralize diplopia for any extended period of time.

Keywords: Strabismus, Internuclear ophthalmoplegia, Wall-eyed internuclear ophthalmoplegia, INO, BINO, WEBINO, Exotropia.

INTRODUCTION

An internuclear ophthalmoplegia (INO) is caused by damage to the medial longitudinal fasciculus (MLF), a structure that connects the 6th and 3rd cranial nerve nuclei to allow for conjugate horizontal eye movements. INO can be unilateral, or bilateral, resulting in a weakening, or loss of adduction of the ipsilateral eye [1]. This is also typically associated with a skew deviation or vertical nystagmus due to interruption of otolithic pathways in the vestibular system [2]. INO can result from many different etiologies, including tumors, infection, hydrocephalus, nutritional, metabolic, and vascular disease; most commonly demyelinating disease, and stroke for unilateral cases [3]. Nonetheless, convergence is frequently spared in these lesions [3]. In a group of thirty patients with INO studied by Kim, he found seventeen men and thirteen women ranging from forty-one to eighty-two years, with a mean age of sixty-two years [3].

In many cases, eyes remain orthophoric with a unilateral INO. Only about 60% of patients develop an exotropia with rather infrequent diplopia [3]. Very few patients have larger angle exotropia known as wall-

eyed bilateral internuclear ophthalmoplegia (WEBINO), though this can also happen in unilateral cases [4].

Several theories exist for the cause of exotropia in WEBINO. An older theory comes from Dr. Martin Lubow, who postulated that the syndrome involved bilateral medial rectus interneurons, or medial rectus subnuclei [5]. This is inconsistent with four cases reported by Chen *et al.* that describe WEBINO caused by lesions at different levels of the brainstem [6]. In three of those cases, the midbrain was involved, but not the medial rectus subnuclei. Chen suggests that the imbalance of vestibular signaling may result from lesions involving the MLF, affecting transmission to the ipsilateral medial rectus, and contralateral eye due to an imbalance of paramedian pontine reticular formation (PPRF) signals. A literature review by Uzawa *et al.* revealed 14 cases of WEBINO caused by stroke, with 6 involving the pons, 5 midbrain, and 3 in both areas [7]. Another theory suggests that this is a result of an unmasked history of controlled exotropia [8]. One final theory from histological studies using dark field micrography suggest that there are small subgroups of medial rectus neurons embedded in the MLF. Damage to these structures may result in an exotropia while leaving the oculomotor nucleus intact [9].

Here, we describe a non-invasive alternative treatment to other forms of management already described in literature. To the best knowledge of the

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authors' knowledge, there have been no publications on convergence exercises for the treatment of WEBINO.

CASE REPORT

A 17-year-old male presented to the eye clinic with a chief complaint of constant horizontal double vision since his craniotomy two months prior. He has an ocular history of myopic astigmatism treated with spectacle wear, with no other significant ocular history.

The patient's medical history is notable for a posterior fossa pilocytic astrocytoma status-post resection and craniotomy on March 3, 2021 (Figure 1). Immediately following the procedure, the patient reported constant horizontal diplopia. The patient and his parents came to the clinic in hopes of scheduling surgery to treat his new onset double vision.

During the clinical exam in May of 2021, the patient was found to have an acuity of 20/20 in the right eye and 20/30 in the left eye with glasses. His pupils were equally reactive without a relative afferent pupillary defect. He had normal intraocular pressure in both eyes. Visual field testing was full via confrontation. Slit lamp exam revealed lagophthalmos of the left eyelid and mild dryness of the cornea. Fundus exam was unremarkable with no optic nerve edema or pallor.

On sensorimotor exam, the patient was found to have a CI-XT strabismus, measuring 8 PD at distance, and up to 20 PD of XT at near, with a right hypertropia (RHT) of 7 PD on primary gaze consistent with a skew deviation. He had minus four limited adduction, minus one limited abduction, and an upbeat nystagmus. His parents were adamant on attempting surgery to help with his diplopia and exotropia. He was able to converge, but with a severe limitation and remote near point of convergence (NPC). To simulate surgery and its possibility of success, a Fresnel prism was applied to his spectacles over his non-dominant eye, with a follow up time dependent on his experience with the prism. Unfortunately, on follow up one week later due to persistent diplopia, it was clear that he was unable to fuse long-term with any amount of prism. The patient and his parents were understandably upset by this finding but agreed to our suggested plan of care.

Since prism therapy was unsuccessful, we recommended against surgery, and instead advised him to try convergence exercises with a progressive strengthening regimen with detailed instructions provided. We explained that since it was still early after neurosurgery, along with his poor ability to converge and the unsuccessful trial of simulated prism, surgery would only provide alignment for one distance, and he would continue to have diplopia anywhere outside of that focal plane. His family was agreeable to this

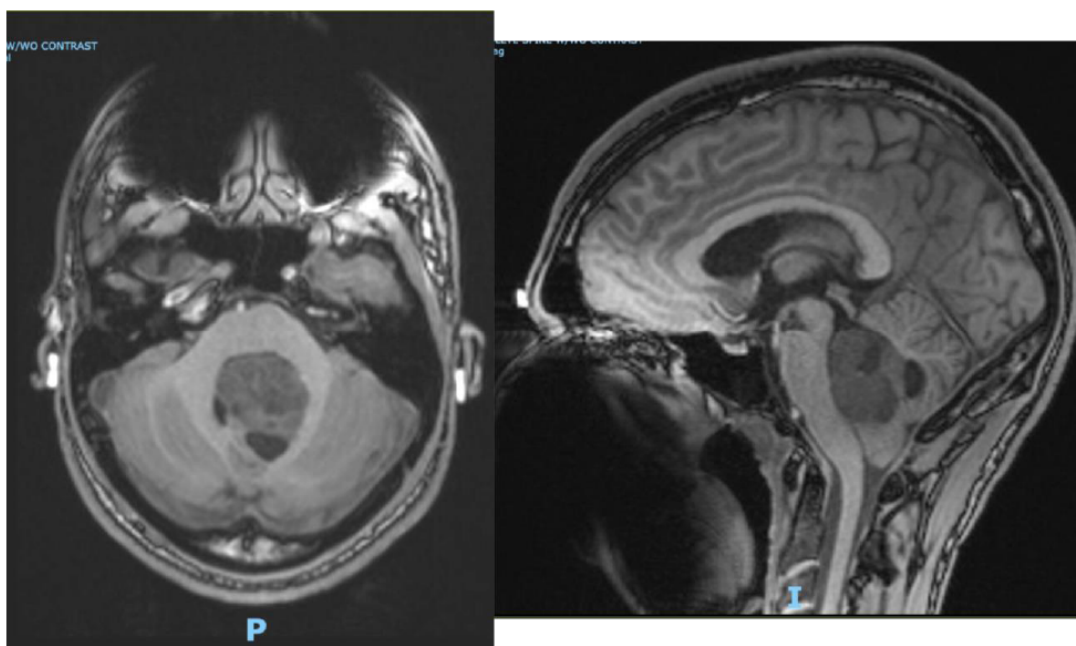


Figure 1: Large solid and cystic fourth ventricular mass with mass effect on the adjacent posterior fossa structures including cerebellar tonsillar descent and crowding of the foramen magnum. The visualized portions of the mass does not restrict diffusion. This is atypical for an ependymoma. The other differential diagnostic consideration is juvenile pilocytic astrocytoma. Similar degree of associated supratentorial hydrocephalus.

alternative plan. The impact of persistent diplopia is significant in a patient who is already significantly affected by problems with coordination. The more we could improve his binocularity, the more likely to benefit his function and quality of life. The decision to do, or avoid surgery, largely depended on his ability to learn to control his eyes over time. Exercises, if successful, would avoid an invasive procedure with the added benefit of allowing the patient to learn how to control his eyes on his own. Surgery done on a patient who cannot fuse may *worsen* function by moving two dissimilar images closer together, making it hard to distinguish the false image.

We continued to follow the patient every three months, and by November 2021, he no longer had any measurable horizontal deviation. Our patient had been consistently doing the recommended convergence exercises. He had a remaining upbeat nystagmus with a RHT of 4 PD at distance and near, mapping out to a skew deviation. This was successfully treated with vertical prism, so a Fresnel was placed onto his spectacles and convergence exercises were continued. By April of 2022, he no longer needed any prism and did not have any further diplopia at distance or near. The patient was able to see comfortably, improving his quality of life, resuming activities such as video games and puzzles. He remained with a BINO, but maintained fusional convergence and was able to eliminate any horizontal strabismus on his own.

DISCUSSION

Management of WEBINO involves eliminating diplopia whether by using occlusion therapy, prism, botulinum toxin injection, or strabismus surgery. Botulinum toxin has demonstrated some effectiveness, though not as much as surgery. Murthy *et al.* reviewed sixteen patients with INO who were treated with injections to one or both lateral rectus muscles [10]. There was symptomatic improvement in the majority of patients, but six still had deviations greater than 20 PD, and only two patients regained some degree of convergence. These patients required an average of 3.5 injections over time. Occlusion therapy and prism are largely considered in smaller-angle strabismus, or post-surgery/injection.

In a small case series by Roper-Hall *et al.*, they identified 8 patients with WEBINO ranging from 25 to 100 prism diopters (PD) of exotropia with the goal of surgery to eliminate diplopia, restore stereopsis, or reduce abnormal head posture [11]. Patients underwent unilateral or bilateral recess-resect

procedures with significant reduction in all patients, with only one patient remaining diplopic. All patients had improved convergence ability and seven patients regained binocularity. Surgery is further supported by another group who treated three patients with WEBINO from multiple sclerosis using bilateral recess-resect procedures [12]. All three patients had resolution of diplopia.

There are no studies or reports of convergence exercises having significant benefit to the treatment of any divergent strabismus in coexisting BINO, while our patient demonstrated success with CI-IXT. Our study has several limitations. This is an isolated case report, so the information may not be generalizable until more studies have been done to identify the rate of success in treating WEBINO with convergence exercises. The patient's improvement could also have been a result of his young age and plasticity of the brain. Exercises may have simply accelerated this improvement, as it is already documented in literature that there is benefit in neurological cases such as post-concussion syndromes [13]. However, it remains to be seen how this modality compares with prism and/or surgical intervention. Larger, randomized-control treatment trials would be beneficial, but difficult given the rarity of this condition.

CONCLUSION

Exact cause and management of WEBINO are still debatable and each patient should be considered on a case-by-case basis. We have successfully demonstrated, at least in one patient, that convergence exercises may be of benefit. Further studies will ideally continue to support this form of management, improve patient care, and continue to clarify when, and if, appropriate.

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Conflicts of Interest

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