Paradoxical Head Tilt in a Congenital Fourth Nerve Palsy

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Neurology® Published Ahead of Print articles have been peer reviewed and accepted for publication. This manuscript will be published in its final form after copyediting, page composition, and review of proofs. Errors that could affect the content may be corrected during these processes.
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David Dongkyung Kim: Drafting/revision of the manuscript for content, including medical writing for content; Study concept or design
John Alexander Fraser: Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Analysis or interpretation of data

Number of characters in title: 56

Abstract Word count:

Word count of main text: 1623

References: 18

Figures: 1

Tables: 0

Supplemental: Patient consent form


Acknowledgements: We thank the patient for providing consent to publish their case and pictures.

Study Funding: The authors report no targeted funding

Disclosures: Amit Kumar Sharma has no conflicts of interest to disclose; David Dongkyung Kim has no conflicts of interest to disclose; John A. Fraser has no conflicts of interest to disclose.
PEARLS

- While most patients with a fourth nerve palsy will adopt a contralateral, “compensatory” head tilt (away from the affected superior oblique) to reduce diplopia, some will instead adopt an ipsilateral, “paradoxical” head tilt (towards the affected superior oblique).
- The Parks-Bielschowsky three-step test remains valid, irrespective of the preferred direction of resting head tilt.
- Hemifacial hypoplasia may be seen ipsilateral to the head tilt and indicates chronicity.
- The superior oblique muscle on the side of a longstanding fourth nerve palsy may appear hypoplastic or atrophic on orbital imaging.
- Early recognition, localization, and management of a congenital fourth nerve palsy in childhood can reduce chronic abnormal head posturing and may avert permanent hemifacial hypoplasia.

OY-STERS

- The direction of head tilt may not always be contralateral to the side of a fourth nerve palsy.
- Some diplopic fourth nerve palsy patients who see two closely spaced competing images prefer the strategy of tolerating more widely spaced diplopia over the effort of maintaining unstable binocular single vision.
A 35-year-old right-handed man was referred for a two-and-a-half-year history of vertical binocular diplopia. His past medical history was notable only for mild young-onset parkinsonism, well-controlled with levodopa-carbidopa. There was no history of trauma. He had a resting head tilt to the right present on old (non-posed) photographs and he affirmed that, throughout his life, barbers and photographers would ask him to straighten up his head. Although his diplopia resolved to single vision with left head tilt, he preferred not to tilt his head this way, as he would develop an uncomfortable feeling of “eye strain” when sustaining this posture. With right head tilt, the two images moved further apart; however, he preferred this position because his eyes felt “more relaxed”. He had previously been prescribed prism glasses, but used them only when it was absolutely necessary to have single vision, as they induced a similar sensation of “eye strain”.

On physical examination, he had normal visual acuity, color vision, funduscopic, and slit lamp examination in each eye. Worth 4-dot testing revealed no suppression. Pupil examination was normal. He had a resting right head tilt, with noticeable right hemifacial hypoplasia (Figure 1A, B). Extraocular motility examination revealed full ductions. Cross-cover examination revealed right hypertropia (4PD (prism diopters)) on primary gaze, unchanged when the patient was laid supine; the hypertropia worsened on left gaze (6PD) and improved on right gaze (3PD). It worsened with right head tilt (16PD) and resolved with left head tilt. The patient was diagnosed with a right fourth nerve palsy with a “paradoxical” ipsilateral head tilt. Consistent with a congenital etiology for this right fourth nerve palsy, he had 12PD of vertical fusional amplitude, and double Maddox rod testing showed no measurable torsion.
General neurological examination was notable only for mild hypomimia, hypophonia, bradykinesia, and rigidity, with the latter two features being greater on the left than the right.

Magnetic resonance imaging (MRI) head revealed both a hypoplastic right superior oblique muscle and underdevelopment of the ipsilateral midface (Figure 1C, D).

Discussion

Fourth nerve palsies are the least common of the ocular motor palsies. They are most often congenital in etiology, accounting for 49% of cases of all fourth nerve palsies. Microvascular disease constitutes the second most common etiology, accounting for an additional 24% of cases. Other, less common etiologies include malignancy and infection.

It is thought that congenital fourth nerve palsies arise either from the absence of the trochlear nerve or from tendon dysfunction of the superior oblique muscle. One radiologic study using thin-section high-resolution MRI showed that, of 97 patients with congenital fourth nerve palsy, 73% had an absent trochlear nerve and atrophy or hypoplasia of the superior oblique muscle. It was presumed that the remainder of cases were due to tendon dysfunction. The peak decades of presentation for congenital fourth nerve palsies are the second and fourth decades of life. Patients with congenital fourth nerve palsies may be asymptomatic until older age, when their binocular fusional reserves diminish in the context of either intercurrent medical illness or age-related extraocular muscle decompensation or remodelling.
A fourth nerve palsy is best diagnosed on physical examination using the Parks-Bielschowsky three-step test, using either alternate cover testing or the Maddox rod to break down binocular fusion and dissociate the two eyes. The three steps include: 1) identifying the eye which is hypertropic in primary position; 2) determining whether this hypertropia increases on right gaze or left gaze; and 3) determining whether the hypertropia worsens with head tilt to the right or left.\(^4\,5\) A fourth nerve palsy produces a hypertropic ipsilateral eye (step 1) whose deviation worsens on contralateral gaze (step 2) and with ipsilateral head tilt (step 3). If desired, prisms can be used to quantify the magnitude of the binocular misalignment in each of the three steps. Because the fourth cranial nerve innervates the superior oblique muscle, which incyclotorts and depresses the eye\(^6\), a patient’s clinical presentation will usually involve an ipsilateral hypertropia causing vertical or oblique diplopia, which may be intermittent. A superimposed torsional component of the diplopia may go unrecognized by the patient or, in the case of a congenital fourth nerve palsy, may be absent altogether. While patients with *acquired* fourth nerve palsies typically have vertical fusional amplitudes similar to those of normal subjects (between 1-3PD), patients with *congenital* fourth nerve palsies can have very large vertical fusional amplitudes, often more than 10PD, and this is a useful clinical test to differentiate between congenital and acquired etiologies for a fourth nerve palsy.\(^7\)

To contend with the diplopia associated with a fourth nerve palsy, many patients subconsciously adopt a resting head tilt.\(^8\) Usually, this head tilt is “compensatory”: a tilt away from the side of the superior oblique palsy, bringing the two images closer together, facilitating binocular image fusion and therefore single vision. Rarely, however, some patients adopt a “paradoxical” head tilt: a tilt towards the side of the superior oblique palsy that further separates the two competing images, thereby allowing one image to be more easily ignored. A paradoxical head tilt results in
greater image divergence: while the ipsilateral superior rectus muscle will activate to elevate and incyclotort the eye, the depressive function of the superior oblique remains impaired, and the summation of these actions causes wider vertical image separation. A paradoxical resting head tilt strategy is preferred in up to 3.4% of those with a fourth nerve palsy (congenital or acquired), potentially due to the constant attention and effort required to sustain single binocular vision via the more common contralateral head tilt.

Patients will often be unaware of their resting head tilt, but old photographs, such as those from family albums or even driver’s licenses, may confirm its chronicity. Spontaneous, non-posed, photographs are the best ones to review, because professional photographers may purposely prompt patients to straighten their heads. When asked directly as part of the history, patients often affirm that hairdressers, barbers, and photographers have, throughout their lives, asked them to straighten up their heads.

Hemifacial hypoplasia is another important clue indicating the longstanding nature of the head tilt (and therefore the fourth nerve palsy). Hemifacial hypoplasia is a consequence of a chronic head tilt that occurs ipsilateral to the tilt – regardless of whether the tilt is “compensatory” or “paradoxical”. Theories of its pathogenesis include chronic changes in internal tissue pressure, low-grade impingement of the carotid artery, or even gravitational effects, with subsequent remodeling of the face over many years. Asymmetries in facial bulk and of the nose, eyes, mouth, and frontalis creases may be seen (as in Figure 1A,B). The finding of hemifacial hypoplasia may be subtle, but the examiner can identify it by mentally connecting the two lateral canthi with a line and the two angles of the mouth with a line and judging whether these two lines are parallel. In our patient, this was evident based on visual observation (Figure 1A, B) and
also seen on MRI (Figure 1C, D). Though hemifacial hypoplasia may begin in infancy, the underlying congenital fourth nerve palsy may not be apparent until later childhood. Early detection and treatment of a congenital fourth nerve palsy in childhood with strabismus surgery can improve and even eliminate chronic abnormal head posturing, with some evidence suggesting this may avert permanent facial asymmetry if undertaken before facial structures mature.\textsuperscript{12–15} The long-term consequences of untreated hemifacial hypoplasia (beyond the cosmetic) are not well studied in the congenital fourth nerve palsy population specifically; however, tooth malposition and jaw malocclusion are known to occur, and are seen also as sequelae of other causes of childhood hemifacial hypoplasia.\textsuperscript{14,16,17}

In our patient, the possibility of cervical dystonia should be entertained in the differential diagnosis, given our patient’s concomitant mild young-onset parkinsonism. However, his head posture is not involuntary, and there is a lack of pain, tremor, or cervical muscular hypertrophy to implicate focal dystonia as a serious diagnostic contender. Similarly, while the combination of parkinsonism and eye movement abnormality may superficially resemble a mitochondrial disorder – a variant of chronic progressive external ophthalmoplegia, for example – the non-progressive nature of our patient’s symptoms, the lack of ptosis, the localization of his ocular motor deficits to just a single cranial nerve distribution, and the lack of family history argue strongly against this.\textsuperscript{18}

Although conventional neurological wisdom is that a longstanding right congenital fourth nerve palsy is accompanied by a compensatory resting left head tilt (and left hemifacial hypoplasia), our case shows that the opposite may occur. The pattern of binocular misalignment during the Parks-Bielschowsky three-step test remains reliable for localization of a fourth nerve palsy:
however, the direction of a patient’s resting head tilt depends instead on their strategy for contending with the diplopia, and the head may not always tilt contralaterally to the affected fourth nerve.
FIGURE 1

Clinical and radiological features of congenital right fourth nerve palsy with chronic paradoxical head tilt. A) Photograph of patient showing a resting right head tilt. B) Features of right hemifacial hypoplasia: lateral canthi and angles of the mouth are not parallel (yellow lines), indicating reduced right facial bulk; shallow nasolabial fold (short white arrow), down-sloping frontalis creases (long blue arrow); larger right nostril, and rightward deviation of the nose are also evident. C) Coronal T1-weighted post-contrast MRI orbits showing hypoplasia of the right superior oblique muscle consistent with a congenital palsy (white arrow); right hemifacial hypoplasia is reflected in the slight rightward deviation of the nasal septum and turbinates (yellow line). D) Right orbit situated inferior to left orbit (white line); rightward deviation of nasal septum (yellow line) consistent with right hemifacial hypoplasia.
## APPENDIX

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<th>Name</th>
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REFERENCES


9. Khan AO. Paradoxical head tilt during fixation with the affected eye in unilateral


doi:10.1097/APO.0b013e31829f4b


doi:10.14219/jada.archive.2001.0055

doi:10.1080/2576117X.2019.1590141

ACKNOWLEDGEMENTS

We thank the patient for providing consent to publish their case and pictures.
"Pearls & Oy-sters:" Paradoxical Head Tilt in a Congenital Fourth Nerve Palsy
Amit Kumar Sharma, David Dongkyung Kim and John Alexander Fraser
Neurology published online April 23, 2021
DOI 10.1212/WNL.0000000000012082

This information is current as of April 23, 2021

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