Cranial Nerve IV: Trochlear Nerve

8

One sentence: The trochlear nerve (Fig. 8.1) is a pure motor nerve, and its lesions result in vertical diplopia, which increases when the gaze is directed downward and medially.

Genetic testing	NCV/ EMG	Laboratory	Imaging	Biopsy
			+	

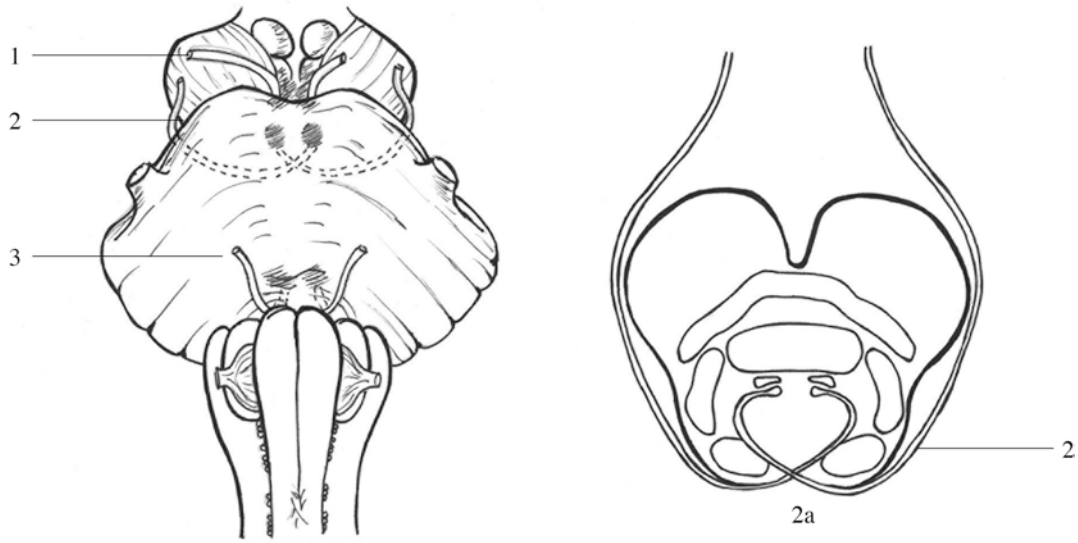

Fig. 8.1 Trochlear nerve. (*1*) Oculomotor nerve, (*2*) *a* fiber crossing, (*3*) abducens nerve

W. Grisold et al., *The Cranial Nerves in Neurology*, https://doi.org/10.1007/978-3-031-43081-7_8

Symptoms

Diplopia: Patients experience vertical diplopia that increases when the gaze is directed downward and medially.

"Torsional diplopia": Trochlear nerve palsy affects the torsion of the eyeball in the plane of the face. Physiologically, the torsion of the eyeball is a normal response to tilting the head sideways.

Signs

The affected eye is sometimes deviated (although this may not be visible to the examiner) and displays less depression during adduction. Hypertropia occurs in severe weakness.

Patients adapt by tilting the head forward to bring the visual fields together. This posture of the head gives a "dejected" appearance ("pathetic nerve" palsies).

Specific Qualities

Motor: +.
Sensory:
Autonomic:
Special senses:
Other:

Location

Central:

Nuclear/fascicular lesion results in a contralateral superior oblique palsy. Lesions distal to the decussation cause an ipsilateral palsy.

Brain stem nuclear or fascicular lesions are not isolated and usually involve the medial longitudinal fascicle (MLF), the sympathetic pathway, or cause afferent pupillary defects (lesion of pretectal fibers).

Central causes include vascular, demyelinating disease, glioma, trauma, and infections.

Intracranial Within the Skull:

The trochlear nerve is the longest intracranial nerve with a length between 60–75 mm, and it decussates before emerging from the brain stem [1].

Lesions occur in the cisternal and subarachnoidal part and also in the cavernous sinus.

Lesions occur due to diabetes, iatrogenic, infection, inflammation, neoplastic, pituitary apoplexy, raised intracranial pressure, and trauma.

Exit of the skull: The nerve exits through the superior orbital fissure outside of the annulus of Zinn.

Outside of the Skull:

Orbital nerve lesions are rare. Signs and symptoms are associated with concomitant lesions of CN II, III, V, and VI. Mechanical restrictions by rheumatoid disease, tendons, trauma.

In orbital lesions proptosis, chemosis and orbital edema are often associated.

Bilateral trochlear nerve lesions present with alternating hypertropia on horizontal gaze or tilt and positive Bielschowsky head tilt test to either side. They are rare, usually observed in trauma [2].

Combination with Other CN

Lesion at the cavernous sinus and in the orbital apex.

Causes and Frequency

Trochlear nerve palsies are well described: [3–6].

Congenital: Rare [7].

Compression: Cavernous sinus, orbital fissure lesions, inflammatory aneurysms (posterior cerebral artery, anterior superior cerebellar artery), tentorial herniation.

Infection: Mastoiditis, meningitis. Herpes zoster [8].

Inflammatory: Ophthalmoplegia or diplopia associated with giant cell arteritis. Local anesthesia [9].

Metabolic: Diabetes.

Myokymia: Superior oblique myokymia.

Neoplastic: Carcinomatous meningitis, cerebellar hemangioblastoma, ependymoma, meningioma, metastasis, neurilemmoma, neurofibroma [2], pineal tumors, trochlear nerve sheath tumors, *e.g.*, schwannoma [10] and others. Orbital apex tumors, cancer metastasis [11].

Pediatric: Congenital, traumatic, and idiopathic.

Trauma: Head trauma causing compression at the tentorial edge, lumbar puncture or spinal anesthesia, subarachnoid hemorrhage, surgery. The trochlear nerve is the most commonly injured CN in head trauma.

Vascular: Arteriosclerosis, diabetes (painless diplopia), hypertension. Rarely in vascular brain stem lesion. Bilateral [12, 13].

Main Investigations

Diagnosis includes clinical optomotor examination and imaging. The diagnosis can be facilitated by the Bielschowsky test.

Suggestive of a trochlear nerve lesion:

Hypertropia of the affected eye.
Diplopia is exacerbated by gazing downward.
Diplopia can be improved by tilting the head away from the affected eye.

Imaging: [14].

Differential diagnosis: Skew deviation, a disparity in the vertical positioning of the eyes of supranuclear origin, can mimic trochlear palsy. Myasthenia gravis, disorders of the extraocular muscles, thyroid disease, and oculomotor palsy that affect the superior rectus can also cause similar signs.

Therapy

The vertical diplopia may be alleviated by the patching of one eye or the use of prisms or sight training. Surgery could be indicated to remove compression or repair trauma.

Prognosis: The recovery rate over a 6-month time period is higher in cases of diabetic etiology than in other nonselected cases.

References

1. Joo W, Rhoton AL Jr. Microsurgical anatomy of the trochlear nerve. Clin Anat. 2015;28(7):857–64.
2. Lekskul A, Wuthisiri W, Tangtammaruk P. The etiologies of isolated fourth cranial nerve palsy: a 10-year review of 158 cases. Int Ophthalmol. 2021;41(10):3437–42.
3. Morillon P, Bremner F. Trochlear nerve palsy. Br J Hosp Med (Lond). 2017;78(3):C38–40.
4. Abkur TM. Trochlear nerve palsy. Pract Neurol. 2017;17(6):474–5.
5. Brazis PW. Palsies of the trochlear nerve: diagnosis and localization—recent concepts. Mayo Clin Proc. 1993;68(5):501–9.
6. Keane JR. Fourth nerve palsy: historical review and study of 215 inpatients. Neurology. 1993;43(12):2439–43.
7. Orphanet. The portal for rare diseases and orphan drugs (Available from: https://www.orpha.net/consor/cgi-bin/OC_Exp.php?lng=EN&Expert=91498).
8. Grimson BS, Glaser JS. Isolated trochlear nerve palsies in herpes zoster ophthalmicus. Arch Ophthalmol. 1978;96(7):1233–5.
9. Chisci G, Chisci C, Chisci V, Chisci E. Ocular complications after posterior superior alveolar nerve block: a case of trochlear nerve palsy. Int J Oral Maxillofac Surg. 2013;42(12):1562–5.
10. Maurice-Williams RS. Isolated schwannoma of the fourth cranial nerve: case report. J Neurol Neurosurg Psychiatry. 1989;52(12):1442–3.
11. Kong E, Koh SA, Kim WJ. Rapid progression from trochlear nerve palsy to orbital apex syndrome as an initial presentation of advanced gastric cancer. Yeungnam Univ J Med. 2019;36(2):159–62.
12. Murray RS, Ajax ET. Bilateral trochlear nerve palsies. A clinicoanatomic correlate. J Clin Neuroophthalmol. 1985;5(1):57–8.
13. Simon S, Sandhu A, Selva D, Crompton JL. Bilateral trochlear nerve palsies following dorsal midbrain haemorrhage. N Z Med J. 2009;122(1300):72–5.
14. Agarwal N, Ahmed AK, Wiggins RH 3rd, McCulley TJ, Kontzialis M, Macedo LL, et al. Segmental imaging of the trochlear nerve: anatomic and pathologic considerations. J Neuroophthalmol. 2021;41(1):e7–e15.