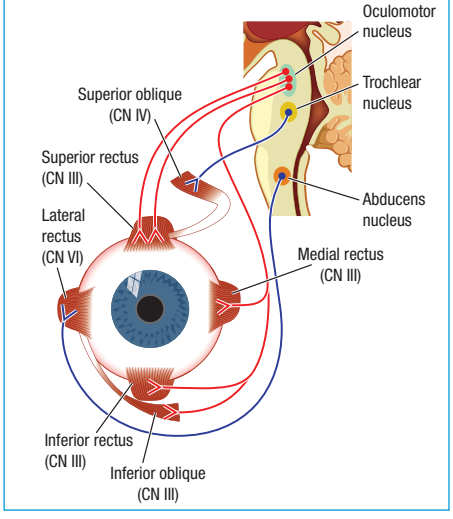
Trochlear nerve palsy

Diplopia is a common presentation at hospital, and the precise cause can be difficult to establish. The commonest cause of vertical diplopia is palsy of the trochlear nerve (CN IV), but signs are subtle and the diagnosis easily missed. This article reviews the neuroanatomy of the trochlear nerve and highlights the features of the history and examination that suggest diplopia is caused by a fourth nerve palsy.

Neuroanatomy

An appreciation of the anatomy surrounding the trochlear nerve is helpful in understanding the clinical presentation of a fourth nerve palsy. The trochlear nerve, containing only somatic motor neurones, is responsible for the innervation of the superior oblique muscle of the eye (*Figure 1*). The effects of this muscle are complex, causing the eye to abduct, depress or intort, depending on the initial starting position of the eye.

Figure 1. Innervation of the extraocular muscles.



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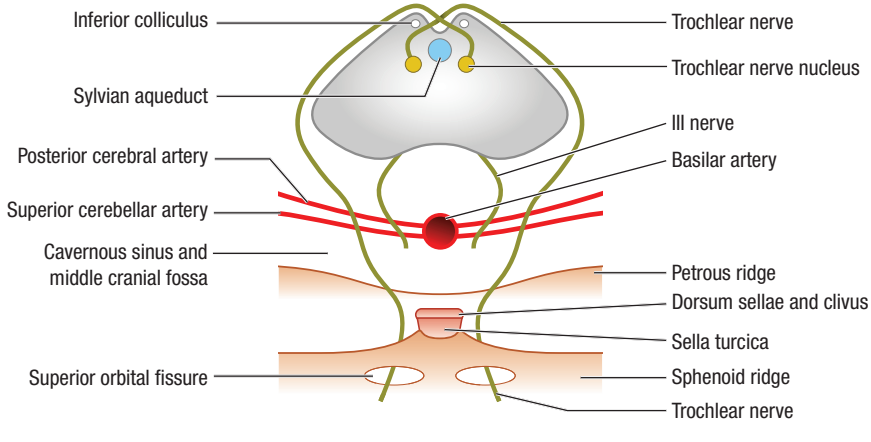
Four special features of the trochlear nerve are of particular clinical relevance. Containing only somatic motor neurones, it is the thinnest and longest of the cranial nerves, and it is the only cranial nerve to emerge from the dorsum of the brainstem (*Figure 2*). These three features make it particularly vulnerable to trauma, especially to the occiput. The final special feature is that is it the only cranial nerve to cross the midline and supply contralateral muscle. The nerve reaches the ventral aspect of the brain by coursing round the cerebral peduncle, eventually joining the oculomotor nerve passing between the superior cerebellar and posterior cerebral arteries, travelling forwards in the lateral wall of the cavernous sinus, through the superior orbital fissure and into the orbit, where it innervates the superior oblique muscle.

Originating posteriorly in the apex of the orbit at the annulus of Zinn, the muscle belly travels anteriorly within the superonasal orbit, with the tendon passing through a cartilaginous ring (the 'trochlea', meaning 'pulley' in Latin). It is then reflected backwards to insert behind the equator of the eye. Contraction of the muscle therefore primarily depresses (especially in adduction), but also serves to intort and abduct, depending on the starting position of the eye. When the right eye is looking to the left, the superior oblique serves to depress the eye, while when it is looking to the right, it applies an intorsional effect.

Figure 2. Course of trochlear nerve.



History

Patients with fourth nerve palsies complain of double vision or images being rotated in relation to each other (*Table 1*). The diplopia is only experienced under binocular viewing conditions (i.e. the second image disappears if either eye is closed) and the images are separated vertically with one image tilted with respect to the other. Patients are usually aware that the double vision is worse when looking in some directions (especially downgaze, when reading or walking down stairs, for instance), but improves or even resolves when looking in other directions; some may resort to closing one eye while reading. The strain of seeing double may also cause headaches. It is always important to ask if there are any other neurological symptoms or health issues, and also whether there is a history of head injury, such as sports-related trauma.

Examination

For the fourth nerve examination, patients should be asked to remove their glasses and be sitting opposite the clinician's chair,

Table 1. Most helpful symptoms and signs for diagnosing a fourth nerve palsy

Symptoms	Torsional diplopia
	Diplopia worse on downgaze (especially walking down stairs and reading)
Signs	Head is tilted away from the lesion when 'looking straight ahead'
	Inferior oblique over-action when looking away from the lesion
	Vertical deviation increases when head is tilted towards the lesion

Table 2. Cover test

This test is useful for determining the presence of strabismus (ocular misalignment).

- For tropia (manifest strabismus), an opaque occluder is held in front of the fixating eye, and the contralateral eye is examined for any movement to take up fixation
- For phoria (latent strabismus) a translucent occluder is used over one eye, and this eye is observed for any movement away from fixation. It will then move back to refixate the target as soon as the eye is uncovered

approximately 1 metre away. First, the eye and periocular area should be inspected, and then ocular movements examined. Finally, the Parks–Bielschowsky three-step test should be performed.

Inspection

This is a crucial stage of the examination which must not be rushed, as it is common to miss key signs at this point.

1. Inspect to see if the head is tilted (ocular torticollis)
2. Inspect the position of the eyes. Are they aligned or vertically separated? A pen torch can help here – with the patient looking at the pen torch, carefully observe the reflection of the light on the cornea. The cover test can also be useful (*Table 2*).
3. Inspect the position of the eyes within the orbit – is there any proptosis?
4. Inspect the eyelids – are there any signs of ptosis or retraction?
5. Inspect the size of the pupils – is there any anisocoria?

In a fourth nerve palsy, the first two features may be abnormal, while the latter three should all be normal. If the head is tilted, it is in the opposite direction to the weak muscle (i.e. tilted away from the lesion). The non-deviating eye has the reflection of the light in its centre, while the deviating eye will have the reflection away from the centre. It is worth also noting that patients can present with their chin facing downwards and eyes deviated upwards.

Ocular movements

Holding one finger vertically (approximately 50 cm from the patient's face), ask the patient to follow your finger with his/her eyes, while keeping his/her head still – you can gently place your hand on the patient's forehead if this is helpful.

Trace the letter 'H' with your finger, ensuring the patient's eyes move to the maximum of their movement, without it becoming too uncomfortable for the patient. The white of the sclera should not be visible on horizontal movement. Observe the eyes to see if one or both eyes do not follow smoothly, or if there is any difference between the eye movements. In a fourth nerve palsy, there is defective depression of the adducted eye, and in horizontal gaze away from the lesion the eye 'scoops upwards' because of the unopposed action of the antagonist muscle (the inferior oblique) (*Figure 3*). Ask the patient if he/she has double vision at any point during this assessment. If so, it is helpful to ask the patient to tell you if the double vision is getting worse or better as you move his/her eyes into various directions of gaze. In a fourth nerve palsy, the double vision will get worse when looking to the side opposite the lesion (e.g. patients with a left fourth nerve palsy experience double vision when looking to the right) and better when looking to the same side. Encourage the patient to describe the image separation as horizontal, vertical, tilted or a combination.

Parks–Bielschowsky three-step test

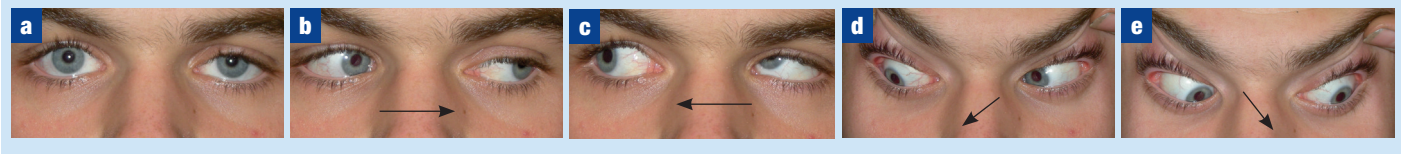
The Parks–Bielschowsky three-step test is an algorithmic assessment used to identify which muscle is weak in patients with vertical diplopia. The three key questions to ask are as follows:

1. Which eye is hypertropic in the primary position? In a fourth nerve palsy, the ipsilateral eye would be higher.
2. Does the hypertropia increase in left or right gaze? Deviation would worsen when looking to the opposite side in a trochlear nerve palsy.
3. Does the hypertropia increase with left or right head tilt? In a fourth nerve palsy, hypertropia increases when tilting to the same side.

The sensitivity and specificity of this test are debatable, with some studies suggesting sensitivity of only 24% in bilateral disease (Muthusamy et al, 2014), and 70% sensitivity in unilateral disease, where all three elements of the test were positive (Manchandia and Demer, 2014). Often, only two of the three elements may be positive in a patient with a fourth nerve palsy, and it is worth noting that no clinical test is 100% sensitive.

Any examination of the fourth nerve is not complete without a full cranial nerve examination and any other necessary neurological or ophthalmological examinations.

Figure 3. Congenital left fourth nerve palsy in a 19-year-old man. **a.** In primary position he fixes with his right eye and his left eye is deviated upwards (hypertropia). **b.** When he looks to the left this vertical deviation disappears, but **(c)** when he looks to the right it is worse as a result of the unopposed action of the antagonist muscle (the inferior oblique). Underaction of the weak left superior oblique muscle is easily seen **(d)** when the patient looks down and to the right in contrast to **(e)** the normal strength of the right superior oblique muscle when looking down and to the left.



KEY POINTS

- The commonest cause of vertical diplopia is a trochlear nerve palsy.
- Palsy of the trochlear nerve is easy to miss, but can be noticed on thorough examination
- Examination of the ocular movements and performing the Parks–Bielschowsky three-step test are useful in diagnosing a fourth nerve palsy.
- Trauma is the commonest cause of a trochlear nerve palsy, as the fourth nerve is the longest and thinnest of the cranial nerves, and because it emerges from the dorsum of the brainstem.
- An appreciation of the differential diagnoses is key, and fourth nerve examination should always be undertaken alongside a full neurological history and examination.

Clinical relevance

Causes of a trochlear nerve palsy are manifold. The three most common causes are traumatic injuries, ischaemic events and congenital disorders. Trauma is the commonest cause (especially to the occiput) because of the length and slender nature of the nerve, as well as its emergence from the dorsum of the brainstem (Akagi et al, 2008; Lyons et al, 2015). Second, ischaemic or vascular disease can cause CN IV palsies as a result of microvascular damage associated with diabetes mellitus, hypertension or vasculitis. Congenital causes, such as maldevelopment of the trochlear nerve, may not present in childhood if adaptations such as a compensatory head posture mask the problem; subsequent decompensation may occur later in life at times of stress, fatigue or intercurrent illness, meaning that patients with congenital abnormalities can still present acutely. The congenital nature of the problem can be revealed by spotting the tell-tale head tilt in old photographs (Akagi et al, 2008). Finally, compressive lesions (intracranial tumours) are a rarer cause, but should be considered for patients in whom there has been no spontaneous recovery after 3 months.

It is important to note that fourth nerve palsies can also be bilateral, usually as a result of a head injury to the occiput. In addition, remember that fourth nerve palsies may not be isolated; combined unilateral palsies of the third, fourth and sixth cranial nerves can occur, causing a 'fixed (unresponsive) dilated pupil', ptosis and ophthalmoplegia. These palsies are caused by lesions in the orbit (e.g. the 'superior orbital fissure syndrome') or in the cavernous sinus. It is therefore important to examine for and consider other neurological deficits which may help to localize the lesion.

When a third nerve palsy is also present, the eye cannot adduct (medial rectus) or elevate (inferior oblique), so there may be no hyperdeviation of the ipsilateral eye associated with the fourth nerve palsy. In some cases of third nerve palsy, conjunctival vessel intorsion when attempting to depress the adducted eye may be visible, a subtle sign that confirms that the fourth nerve is intact. However, in most cases of third nerve palsy, the ophthalmoplegia is so profound that it is not possible to say if the fourth nerve is involved or not.

Differential diagnoses

Although a fourth nerve palsy is the commonest cause of vertical diplopia, other causes must also be considered.

First, brainstem disease resulting in skew deviation can cause diplopia, where double vision disappears when the patient is lying flat and looking upwards towards the ceiling. However, this pattern of disease will not conform to the examination findings for the Parks–Bielschowsky three-step test as outlined previously (Wong, 2010). Second, neuromuscular junction disorders can occur, a good example of which is myasthenia gravis, commonly presenting with a history of image separation varying from day to day, and ptosis on examination. Third, orbital disorders such as thyroid eye disease can present with diplopia. It is important to look for other symptoms in the history of hyperthyroidism, and to ask about any previous orbital or eye muscle surgery, even if it took place a long time ago.

Investigation

In terms of investigating a patient with signs and symptoms of a fourth nerve palsy, all potential causes should be taken in to account. For instance, myasthenic serology (acetylcholine receptor and muscle kinase antibodies), thyroid status tests (triiodothyronine, thyroxine, thyroid-stimulating hormone and thyroid antibodies) and vascular status tests (blood glucose and lipids, erythrocyte sedimentation rate and C-reactive protein) may be relevant in some patients. Ultimately, however, magnetic resonance imaging with contrast enhancement is recommended for most patients with suspected trochlear nerve palsy (Tamhankar and Volpe, 2015).

Conclusions

When presented with a patient with vertical diplopia, the most common cause is a fourth nerve palsy, and this must be considered while taking a history and examining the patient. Signs can be subtle and are often missed, making the diagnosis more difficult to establish than that of a third or sixth cranial nerve palsy. Remember that fourth nerve palsies do not always occur in isolation, and so a full neurological history and examination must always be performed. **BJHM**

Conflict of interest: none.

Akagi T, Miyamoto K, Kashii S, Yoshimura N (2008) Cause and prognosis of neurologically isolated third, fourth, or sixth cranial nerve dysfunction in cases of oculomotor palsy. *Jpn J Ophthalmol* **52**(1): 32–35.

Lyons C, Godoy F, AlQahtani E (2015) Cranial nerve palsies in childhood. *Eye (Lond)* **29**(2): 246–251. https://doi.org/10.1038/eye.2014.292

Manchandia AM, Demer JL (2014) Sensitivity of the three-step test in diagnosis of superior oblique palsy. *J AAPOS* **18**(6): 567–571. https://doi.org/10.1016/j.jaapos.2014.08.007

Muthusamy B, Irsch K, Peggy Chang HY, Guyton DL (2014) The Sensitivity of the Bielschowsky Head-tilt test in diagnosing acquired bilateral superior oblique paresis. *Am J Ophthalmol* **157**(4): 901–907. https://doi.org/10.1016/j.ajo.2014.01.003

Tamhankar MA, Volpe NJ (2015) Management of acute cranial nerve 3, 4 and 6 palsies: role of neuroimaging. *Curr Opin Ophthalmol* **26**(6): 464–468. https://doi.org/10.1097/ICU.0000000000000200

Wong AM (2010) Understanding skew deviation and a new clinical test to differentiate it from trochlear nerve palsy. *J AAPOS* **14**(1): 61–67. https://doi.org/10.1016/j.jaapos.2009.11.019

Further reading

Bowling B (2016) *Kanski's Clinical Ophthalmology*. 8th edn. Elsevier, London

Denniston A, Murray P (2014) *Oxford Handbook of Ophthalmology*. 3rd edn. Oxford University Press, Oxford

Kalra P (2014) *Essential Revision Notes for MRCP*, PasTest Ltd, Cheshire

Kline L (2014) *Neuro-Ophthalmology - Basic and Clinical Science Course – Section 5*. American Academy of Ophthalmology, Canada

Park UC, Kim SJ, Hwang JM, Yu YS (2008) Clinical Features and natural history of acquired third, fourth, and sixth cranial nerve palsy. *Eye* **22**(5): 691–696. https://doi.org/10.1038/sj.eye.6702720

Riordan-Eva P, Whitcher J (2007) *Vaughan and Asbury's General Ophthalmology*. 17th edn. Lange, New York