A simple guide to nystagmus

Christian J Lueck

Nystagmus is a potentially mind-boggling subject, as over forty different types are recognized. However, by classifying the various types into a small number of categories, the clinician can make sense of them, and so make use of a very important clinical sign.

Nystagmus refers to rapid 'to-and-fro' movement of the eyes. It is a normal phenomenon in certain situations, e.g. looking out of a moving railway carriage (optokinetic nystagmus) or when on a merry-go-round (vestibular nystagmus). There are, however, many types of nystagmus which occur as manifestations of neurological disease and some of these are more serious than others. The ability to recognize the various different types of nystagmus is an extremely useful clinical skill.

This article sets out to try to simplify what is potentially a mind-boggling subject. More detailed descriptions can be found in Leigh and Averbuch-Heller (1998), Leigh and Zee (1999), or Burde et al (1992), and a detailed guide to the physiology of eye movements and nystagmus can be found in Carpenter (1988).

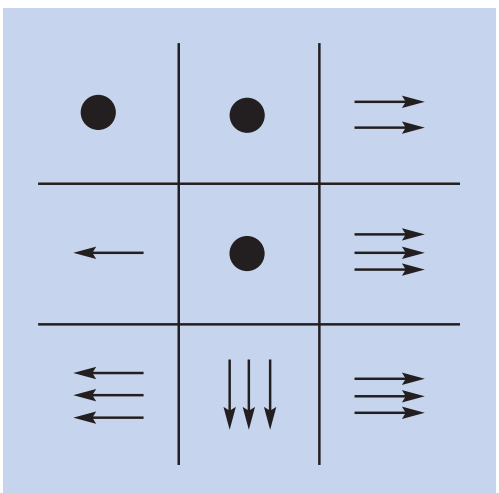

Figure 1. Example of nystagmus documentation in the nine cardinal positions of gaze. The fast phase direction is shown by the direction of the arrow (pendular nystagmus is shown as a double-headed arrow), beat amplitude by the length of the arrow (usually three different arbitrary lengths), and beat frequency by the number of arrows (1–3).

HOW TO EXAMINE THE PATIENT AND DOCUMENT IT

First a word about examination. The patient must be examined for nystagmus in each of the nine cardinal positions of gaze. In each position, the nystagmus must be analysed for direction, amplitude and beat frequency. These can be represented in a diagram (*Figure 1*) which allows documentation and comparison over time. If the two eyes move disconjugately, two separate diagrams must be used.

HOW TO CLASSIFY NYSTAGMUS

Simple classifications are possible in terms of either direction or waveform of the nystagmus. The direction of the fast phase can be up, down, right, left or torsional. This is not much help if the nystagmus is conjugate (i.e. both eyes are doing the same thing), but is useful if it is disconjugate. Horizontally disconjugate movements occur in convergence-retraction nystagmus (CRN), and vertically disconjugate movements in see-saw nystagmus (SSN). Both are discussed further below. 'Ataxic' nystagmus (i.e. nystagmus of the abducting eye) is a manifestation of an internuclear ophthalmoplegia, and should be treated as such.

Nystagmus waveform is sometimes useful if it can be determined. Nystagmus can be categorized into three types: 'jerk' (fast and slow phases), 'pendular' (no obvious fast phase) and 'complex' (*Figure 2*). Jerk nystagmus is subclassified on the basis of the slow phase waveform, which may be linear (typical of vestibular nystagmus), exponentially decreasing (typical

Dr Christian J Lueck is Consultant Neurologist and Part-Time Senior Lecturer in the Department of Clinical Neuroscience, Western General Hospital, Edinburgh EH4 2XU

of cerebellar/brainstem nystagmus), or exponentially increasing (pathognomonic of congenital nystagmus). Complex waveforms are usually manifestations of congenital nystagmus (*Figure 2*). It can be extremely difficult to distinguish these clinically, so this classification is not much use unless sophisticated eye movement recording equipment is available. However, the distinction between jerk and pendular waveforms is useful, and this should be detectable clinically as there is no fast phase in the latter. Pendular nystagmus is often seen in the context of congenital nystagmus, but may occur as an acquired phenomenon, usually in diseases of myelin (e.g. multiple sclerosis), brainstem stroke or, rarely, in the context of oculopalatal myoclonus.

Nystagmus has also been classified alphabetically and in terms of aetiology. Neither of these is useful clinically as they put the cart before the horse.

Practically speaking, the only clinically useful classification of which the author is aware is based on clinical frequency (*Table 1*). There are three common types of nystagmus, and three less common (but clinically important) types. Most of the remainder are extremely rare 'collector's items'. However, it is important to be able to recognize a few additional conditions which resemble nystagmus, as this can often prevent unnecessary/inappropriate investigation of patients.

COMMON TYPES OF NYSTAGMUS

Vestibular nystagmus

This can, of course, be physiological. When pathological, the nystagmus can arise as a result of damage to the peripheral vestibular apparatus/vestibular nerve, or damage to the central vestibular pathways. If caused by a peripheral lesion, horizontal nystagmus is directed to the side opposite the lesion, and is often more pronounced when the patient looks into the direction of the fast phase (Alexander's law).

Vertical and torsional nystagmus can also occur as a result of peripheral mechanisms, but not pure vertical nystagmus in the midline — this implies a central lesion. A combination of pure vertical nystagmus when looking to the right and pure torsional nystagmus when looking to the left (or vice versa) implies damage to a vertical semicircular canal. Peripheral vestibular nystagmus is usually transient, settling after at most 2 weeks, as it can be suppressed by vision. Removing visual fixation (e.g. in the dark or by using Frenzel spectacles) may bring out suppressed nystagmus of this sort.

Vestibular nystagmus as a result of central mechanisms is usually persistent, and not suppressed by vision. The direction of the nystag-

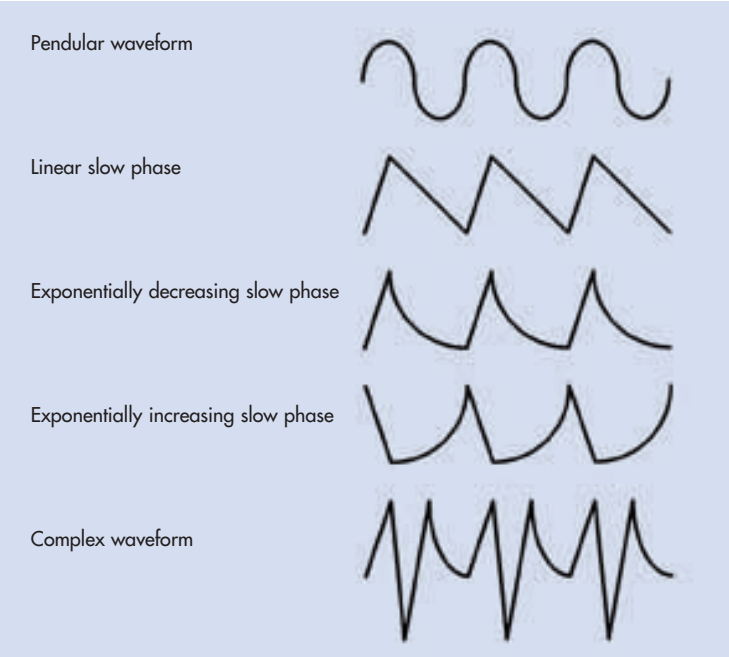

Figure 2. Diagram to show the different types of nystagmus waveform: pendular, jerk and complex, with jerk nystagmus being further subdivided into linear, exponentially increasing and exponentially decreasing waveforms.

TABLE 1.
Practical classification of nystagmus

Types of nystagmus	Common	Vestibular disease
		Cerebellar/brainstem disease
		Congenital nystagmus
	Uncommon but important	Periodic alternating nystagmus
		See-saw nystagmus
		Convergence-retraction nystagmus
	Rare	Epileptic nystagmus
		Eyelid nystagmus
		Pursuit nystagmus
Conditions resembling nystagmus	Disorders of saccadic function	Square wave jerks
		Macro-square wave jerks
		Ocular flutter
		Opsoclonus
		Voluntary nystagmus
	Other entities	Superior oblique myokymia
		Ocular bobbing
		Ping-pong gaze
		Oculopalatal myoclonus
		Oculomasticatory myoarrhythmia
		Spasmus nutans

mus may be affected by head position (because of the effects of gravity).

Cerebellar/brainstem nystagmus

The most common type, gaze-evoked nystagmus, arises as a result of inadequate gaze-holding mechanisms, allowing drift of the eyes back to primary position, with subsequent fast phases to pick up fixation again. The direction of the nystagmus is therefore dependent on the direction of gaze. Typically, unilateral cerebellar lesions cause nystagmus when the eyes are looking towards the side of the lesion, but this is not always so.

Other types of nystagmus may be seen in cerebellar/brainstem disease, including periodic alternating nystagmus (PAN) and rebound nystagmus (transient nystagmus in primary position following prolonged eccentric fixation). There may also be other eye movement disorders such as macro-square wave jerks, or saccadic hypermetria (see below).

Congenital nystagmus

This is not infrequent. It usually presents in infancy or childhood, but may present to adult neurologists, or be seen as an incidental finding, and thereby cause unnecessary alarm/excess investigation if not correctly diagnosed. The nystagmus is usually horizontal, and there is often a 'null position' in which no nystagmus is evident. This is not necessarily straight ahead, and patients may develop head tilts or turns, and may also exhibit head nodding.

The nystagmus is typically worse with fixation, attention or anxiety, but is not affected by head posture. It often improves on convergence or eye closure. There are many possible waveforms, including pendular, jerk and complex. Exponentially increasing slow phases are pathognomonic, but this may be difficult to assess clinically. There may be associated clinical signs which are pathognomonic, including reversal of smooth pursuit or optokinetic nystagmus, latent or 'flash' nystagmus (conjugate nystagmus brought on by covering one eye/shining a bright light into one eye), and alternating sursumduction (elevation of a covered eye — note that this must occur bilaterally to distinguish it from a hypertropic squint).

TABLE 2. Causes of periodic alternating nystagmus

Cerebellar disease	Multiple sclerosis
	Trauma
	Tumour
	Degeneration
Brainstem disease	Infarction
	Arnold–Chiari malformation
Congenital nystagmus	

INFREQUENT BUT IMPORTANT TYPES OF NYSTAGMUS

Periodic alternating nystagmus

In PAN there is a slow alteration of nystagmus direction, almost always in the horizontal plane: initially right-beating, the amplitude gradually declines to zero, and then resumes, this time left-beating. This increases in amplitude to a maximum, and then declines again, through zero and back to right-beating. The whole cycle usually takes about 4–5 minutes, so can be missed by the unwary. Rarely, the vertical plane may be involved as well, in which case this is known as 'windmill' nystagmus.

PAN can be seen in the context of a number of conditions listed in *Table 2*; it is sometimes precipitated by visual loss (such as intraocular haemorrhage or cataract). Importantly, PAN is usually responsive to treatment with baclofen, and therefore should not be missed.

Convergence-retraction nystagmus

Also known as the Körber–Salus–Elschnig syndrome, CRN is usually seen in the context of dorsal midbrain lesions. All extraocular muscles co-contract simultaneously, and, because the medial recti are the strongest muscles, there is net convergence. This phenomenon is most easily elicited by asking the patient to look up, or by using an optokinetic drum with stripes moving downwards. Because of the possibility of pinealoma or other neurosurgically treatable lesion, CRN is also important to recognize and investigate appropriately.

See-saw nystagmus

In SSN, the eyes move disconjugately in the vertical plane, accompanied by a degree of torsion (*Figure 3*). It is most easily detected clinically by focussing on a point midway between the patient's eyes. It can occur congenitally, particularly in the context of retinitis pigmentosa, albinism, septo-optic dysplasia, or Arnold–Chiari malformation. However, it may also be seen as an acquired phenomenon, typically with parasellar masses, brainstem stroke, syringobulbia or head trauma. It is less easy to treat than PAN, but does sometimes respond to baclofen, clonazepam or alcohol.

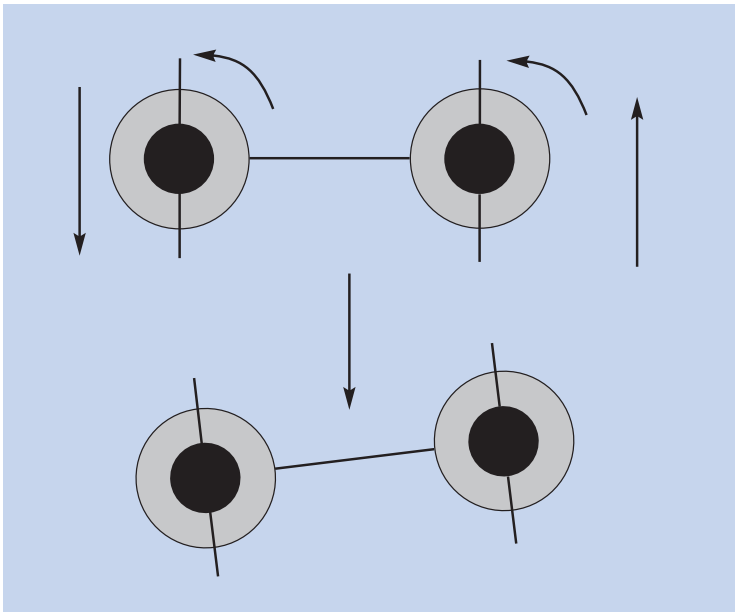

Figure 3. Diagram to show the eye movements in see-saw nystagmus. The elevating eye (in this case the left) intorts, while the other eye extorts.

RARE TYPES OF NYSTAGMUS

There are a few extremely rare types of nystagmus which are of little clinical relevance, including epileptic nystagmus, eyelid nystagmus and pursuit nystagmus.

CONDITIONS RESEMBLING NYSTAGMUS

Disorders of saccadic function

Square wave jerks/macro-square wave jerks: In some normal subjects, small saccades occur which temporarily take the eyes off target, being followed by a refixation saccade after 200 ms (the normal minimum intersaccadic interval) (*Figure 4*). These are therefore not necessarily pathological, but they are seen in increased frequency in degenerative conditions such as progressive supranuclear palsy, Alzheimer's and motor neurone disease, and are also more common in patients with schizophrenia. They can be confused with nystagmus if they occur frequently.

If the amplitude of the jerks is larger than 7° (i.e. clinically very obvious), they are referred to as 'macro-square wave jerks': these are pathognomonic of cerebellar disease.

Ocular flutter/opsoclonus: If the normal intersaccadic interval does not occur, 'back-to-back' saccades may be seen. These are most commonly restricted to the horizontal plane — ocular flutter (*Figure 4*). If the saccades occur in all directions, this is referred to as opsoclonus (or 'saccadomania'). This is seen as part of the opsoclonus–myoclonus syndrome, thought to be viral in aetiology, but may also be a paraneoplastic phenomenon, particularly in association with neuroblastoma in children, or breast cancer in adults (when it may be associated with anti-Ri antibodies). The pathophysiology is thought to involve a failure of 'pause cell' function in the brainstem. There is no specific treatment, but spontaneous recovery usually occurs in postviral or other infective causes.

Voluntary nystagmus: Some patients are able to generate a series of back-to-back saccades by

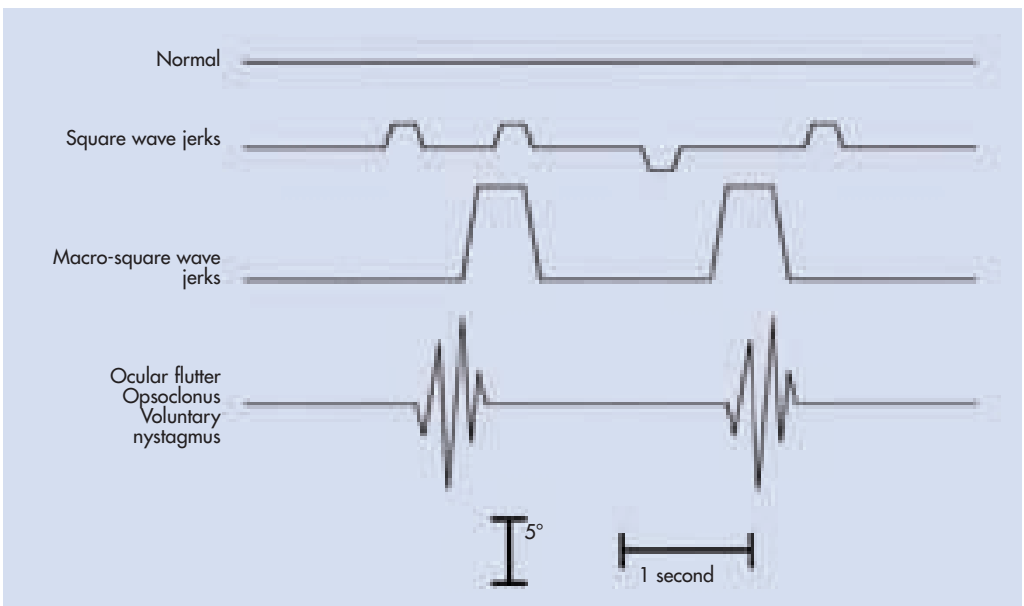

Figure 4. Diagram to show the various types of saccadic abnormality which may be mistaken for nystagmus. Time is represented on the horizontal axis, and saccades (fast eye movements) represent movement to right or left (vertical axis).

slightly converging and defocussing their eyes. This generates 'bursts' of nystagmus which can fool the unwary. It is not associated with underlying organic pathology and early recognition will prevent unnecessary or inappropriate investigation.

Other entities resembling nystagmus

Superior oblique myokymia: This usually presents as (torsional) oscillopsia. It is generally benign, although occasionally is the harbinger of brainstem disease such as a glioma. Membrane stabilizing agents such as carbamazepine are often useful in suppressing it.

Ocular bobbing/ping-pong gaze: These are seen in disorders of the posterior fossa (especially pons and cerebellum), including haemorrhage and trauma, but also in metabolic/toxic encephalopathy. They are usually seen in the context of comatose patients on the intensive care unit. Ocular bobbing comprises rapid downward movements from primary position followed by slow return. Depending on whether the first movement is down or up, fast or slow, various permutations have been described as 'inverse ocular bobbing', 'reverse ocular bobbing', and 'converse ocular bobbing'. Ping-pong gaze refers to the slow alteration of gaze from right to left and back again, typically seen in posterior fossa haemorrhage, but also in bilateral cerebral infarction. It is associated with an extremely poor prognosis.

Oculopalatal myoclonus/oculomasticatory myoarrhythmia: These are rare disorders. A lesion of the part of the brainstem between the red nucleus, inferior olive and dentate nucleus (the Guillain–Mollaret triangle), typically a stroke, may result in a combination of palatal myoclonus and vertical pendular nystagmus, so-called oculopalatal myoclonus. There is, unfortunately, no satisfactory treatment for this condition which may be quite disabling. Oculomasticatory myoarrhythmia involves pendular convergent movements of the eyes with co-contraction of the masseter muscles, and is said to be pathognomonic of cerebral Whipple's disease.

Spasmus nutans: This comprises the triad of head nodding, nystagmoid eye movements, and anomalous head postures. It is seen in children between the ages of 1 and 8 years, and is totally benign. If correctly diagnosed, the patient can be spared cerebral imaging (which would require an anaesthetic in this age group).

CONCLUSIONS

There are three common types of nystagmus which should be identified: vestibular, cerebellar/brainstem and congenital. It is useful to be able to recognize a further group of three less common types to allow appropriate management: PAN, CRN and SSN. Recognition of the conditions which mimic but are not nystagmus (e.g. saccadic disorders) will also allow more appropriate management. Hopefully, this classification provides a framework upon which to 'peg' the various types of repetitive eye movements which are encountered in clinical practice, thereby simplifying what might otherwise be a very complex subject. HM

Conflict of interest: none.

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KEY POINTS

- There are three common types of nystagmus: vestibular, cerebellar/brainstem and congenital.
- Pure vertical nystagmus in primary position is never caused by peripheral vestibular disease.
- Periodic alternating nystagmus (PAN), see-saw nystagmus (SSN) and convergence-retraction nystagmus (CRN) are uncommon but important and often treatable.
- It may take several minutes of observation to detect PAN.
- The easiest way to see SSN is to look at a point equidistant between the eyes.
- CRN is best elicited by asking the patient to look up, or using a downwardly rotating optokinetic drum.
- Many conditions, particularly saccadic disorders can be confused with nystagmus. Recognition of these conditions can sometimes prevent unnecessary/inappropriate investigation.
- A simple method of documentation is described.