Nystagmus

Christian J. Lueck
Associate Professor of Neurology,
Department of Neurology, The Canberra
Hospital, Australia; E-mail: christian.lueck
@act.gov.au
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INTRODUCTION
Most readers will already know that nystagmus
is a to-and-fro movement of the eyes. Because it
is a normal as well as an abnormal phenomenon,
human beings must have been aware of it for
many millennia. However, the first formal de-
scription of the oculomotor response to vestibu-
lar stimulation is credited to Erasmus Darwin in
1796, and of optokinetic nystagmus to Purkinje
in 1819. The term is apparently thought to de-
rive from the jerky head-nodding which occurs
when a person drifts off to sleep in the upright
position.

Nystagmus can occur in any direction – hori-
zontal, vertical or torsional – and it has several
different possible waveforms (Fig. 1). Most doc-
tors should be able to distinguish jerk nystagmus
from pendular and complex waveforms, though
this can sometimes be difficult. Jerk nystagmus
is the most common type: there is a fast saccadic,
usually corrective, phase interspersed between
slow movements (usually pathological) in the opposite direction. There are, in fact, three possible waveforms for the slow phase of jerk nystagmus (Fig. 1), but these are usually extremely difficult to distinguish with the naked eye, and such subdivision is therefore not clinically useful.

**DOCUMENTATION OF NYSTAGMUS**

It is important to be able to document a patient’s nystagmus quickly and accurately not just to help in the diagnosis of where and what the lesion might be, but also for comparison over time, and to ease communication. The best way to do this is to draw up a 3 × 3 grid (Fig. 2). Each of the nine boxes represents one of the nine cardinal positions of gaze (primary position, right, up and right, up, up and left, etc.). The direction of the arrows represents the direction of the fast phase which may be torsional, or even a combination of directions (e.g. right-beating with added right torsional nystagmus). If the waveform is pendular, double-headed arrows are used. The number of arrows gives an estimate of the frequency of the nystagmus, while the length indicates the amplitude. If the nystagmus is disconjugate for any reason, two grids will be needed – one for each eye.

**CLINICAL ASSESSMENT OF NYSTAGMUS**

In Medline and the Cochrane database there are almost no formal analyses of nystagmus in terms of sensitivity, specificity, positive predic-
Is it pathological?
This is not usually a problem because the two physiological stimuli that induce ‘normal’ nystagmus – vestibular stimulation and optokinetic stimulation – are usually obvious. However, there are a couple of potential traps:

• ‘End-point’ or ‘physiological’ nystagmus is seen in about 5% of subjects. This is horizontal nystagmus only on looking to the far right or left. This will not be a problem if extremes of gaze and excessive convergence are avoided when testing eye movements.

• ‘Voluntary’ nystagmus is something that most people can generate with practice. Some individuals become particularly expert and use it as a party trick, but it can be seen in patients with a variety of somatoform disorders. The movement in fact comprises back-to-back saccades without an intersaccadic interval and so is not really true nystagmus (Fig. 1). Nevertheless, it may be interpreted as such by the unwary clinician, and patients may be subjected to many unnecessary tests because this possibility has not been considered.

Is it really nystagmus?
Not all to-and-fro eye movements are nystagmus. The most frequent ‘look-alikes’ are saccadic disorders, but several other types of rather nystagmoid eye movement are given names such as myokymia, myorhythmia, myoclonus, and bobbing. These nystagmoid movements are often of relatively large amplitude and are more irregular than true nystagmus (Leigh et al. 1999).

In many cases, however, the distinction appears to be historical or conventional.

Saccadic abnormalities are, however, sufficiently common to warrant specific comment. Under normal circumstances, saccadic eye movements are used to redirect the line of sight to a target of interest. Nevertheless, spontaneous (involuntary) saccades do occur and, when they occur, the eyes need to be returned to the desired direction of gaze. This deliberate return movement usually has to wait for the obligate intersaccadic interval of 150–200 ms, generating a square waveform (Fig. 1). If the amplitude is small, these movements are known as square wave jerks. They can be seen as a normal phenomenon, particularly in the context of stress, but they have also been reported to occur more frequently in patients affected by, for example, dementia, schizophrenia, progressive supranuclear palsy and AIDS. As such, square wave jerks have low specificity and sensitivity and so are not a useful clinical sign. However, if the amplitude is more than 7° (i.e. very obvious clinically) they are felt to be pathognomonic for cerebellar disease (macro square wave jerks).

It is possible for back-to-back saccades to occur without an intersaccadic interval as in voluntary nystagmus described above. When this occurs pathologically, there are bursts of intrusive back and forth eye movements. If confined to the horizontal plane, this is known as ocular flutter, if they occur in all directions, opsoclonus. Both are usually associated with visual degradation, and often oscillopsia. Assuming that voluntary nystagmus has been eliminated, both flutter and opsoclonus are always pathological, usually the result of cerebellar or brainstem disturbance.

Is it congenital or acquired?
Congenital nystagmus is generally obvious in childhood, but it may occasionally present in adult life, often as an incidental finding. In this situation, it is usually associated with very little, if any, functional visual degradation, and so is asymptomatic. It can, however, cause problems for the neurologist if patients are observed to have nystagmus in the context of unrelated symptoms or other physical signs. Rather like voluntary nystagmus, this can result in inap-
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appropriately and unnecessary investigation and, occasionally, incorrect diagnosis.

Determining that nystagmus is actually congenital in this situation may be quite difficult clinically and, if unsure, it is better to assume that it is pathological. However, there are a number of clues which should alert the clinician to this possibility, particularly if the nystagmus doesn’t seem to fit the rest of the clinical picture. The first is obviously the history as many patients have known about their ‘dancing eyes’ all their lives. Head shaking or nodding may be seen, and are both unusual in acquired nystagmus. Other indications that the phenomenon is congenital include reversal of the direction of optokinetic nystagmus or smooth pursuit. Similarly, being able to elicit nystagmus by covering one eye (latent nystagmus) is pathognomonic for congenital nystagmus – this may first become evident when performing ophthalmoscopy. Finally, eye movement recording can often demonstrate unequivocally that nystagmus is congenital because there are a number of waveforms which are pathognomonic, but this investigation is not always available.

What is the cause?
The most common causes of pathological nystagmus are disturbance of vestibular function, or of the normal gaze-holding mechanisms in the brainstem and cerebellum. There are, however, many other types of nystagmus which may be related to visual loss, or to disturbance of ocular motor control at supranuclear, internuclear, nuclear or infranuclear levels (Leigh & Zee 1999; Leigh & Rucker 2005).

Is it affecting the patient’s vision?
Curiously, not all forms of nystagmus affect visual acuity. Likewise, some forms of nystagmus generate marked movement of the perceived visual world (oscillopsia) while others do not. Why some pathologies result in oscillopsia while others do not is an interesting, but as yet unanswered question. The significance of these points relates to management. For example, downbeat nystagmus associated with marked oscillopsia will usually require attempted symptomatic treatment (e.g. baclofen, carbamazepine or 3,4 diaminopyridine, intramuscular injections of botulinum toxin, spectacle prism corrections, or ocular muscle surgery). If the nystagmus is asymptomatic, there is of course no need to treat the nystagmus itself.

Can it be treated?
This is beyond the scope of this article but is discussed by Leigh & Rucker (2005).

REFERENCES

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practice points
• Nystagmus is a common physical sign, seen in a large variety of ophthalmological and neurological conditions
• However, precise estimates of its true sensitivity and specificity for various diagnoses are not currently available
• With the exception of the ‘catches’ described, nystagmus generally implies some form of underlying pathology and needs to be investigated appropriately
• Nystagmus should be distinguished from nystagmus-like eye movements such as saccadic disorders, myokymia, myorhythmia, myoclonus and bobbing
• Congenital nystagmus should be considered when an incidental finding of nystagmus does not appear to fit the rest of the clinical picture
• The management of nystagmus is strongly influenced by how much visual degradation or oscillopsia it generates