# Management of nystagmus

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## Introduction

The previous article in this two-part series, *Clinical Assessment of Nystagmus*, covered classification and diagnosis of the various forms of nystagmus. This article discusses the options available for managing patients with the condition.

The range of interventions available largely depends on the underlying type of nystagmus and symptoms that are present. This article is therefore divided into therapies for each of the major nystagmus groups discussed in the previous article: *latent nystagmus*, *infantile nystagmus* and *acquired nystagmus*.

### The effects of nystagmus

Regardless of the underlying condition, in most cases, nystagmus causes functional visual difficulties. These difficulties manifest in a number of different ways. For example, the presence of oscillopsia (illusory motion of the world[1]) can be extremely debilitating and disorienting, making visual information difficult to interpret. Individuals with early-onset nystagmus usually have reduced visual acuity (VA), even in idiopathic cases of infantile nystagmus (IN)[2]. In addition to visual disability, nystagmus may be associated with profound social and emotional difficulties[3]. Patients may also wish for a reduction in nystagmus intensity for cosmetic reasons.

The following three steps summarise an 'ideal' management plan for any patient with nystagmus:

1. Treat the underlying condition (if any)
2. Fully correct refractive error
3. Minimise nystagmus intensity

As mentioned in the previous article, all patients should have had a full workup with a hospital eye service before being managed in optometric practice. Most individuals with nystagmus will also benefit from a low vision assessment, and the usual measures such as enlargement of text and use of reading aids are often of great help. Due to the 'slow to see' phenomenon in many forms of nystagmus, it is wise to communicate to parents and teachers the benefits of allowing extra time for schoolwork.

One particularly valuable resource that can be offered to any individual with nystagmus is awareness of the support networks that exist. *Nystagmus Network* is a national charity and support group which can provide families with practical help and information for children as they grow up with the condition, as well as adults newly diagnosed with nystagmus[4]. Patient information leaflets are available from the charity on request.

### Management of latent nystagmus

Latent nystagmus typically presents as part of an infantile strabismus syndrome. Optical (e.g. refractive correction for accommodative esotropia) and/or surgical alignment of the eyes in these cases can reduce nystagmus intensity, convert manifest-latent nystagmus to latent-latent nystagmus and improve binocular VA[5].

The management of amblyopia associated with latent nystagmus deserves special consideration, as latent nystagmus will worsen with traditional occlusion therapy. Blur or atropine therapy should be used instead. Improvements in VA following amblyopia treatment are also associated with permanent reductions in nystagmus intensity[6].

Finally, latent nystagmus is often associated with a significant head turn towards the fixing eye, especially in cases of monocular infantile blindness, in which there is often a coexisting large angle esotropia. Surgery for abnormal head posture (AHP) in latent nystagmus is usually performed on the *dominant* or *fixing* eye. In cases of existing esotropia, an initial combined recess-resect surgery in the fixing eye may be sufficient to correct the AHP, although additional surgery may be needed in some cases[7].

## Management of infantile nystagmus

Once established in early childhood, IN is a lifelong condition. At present, there is no effective 'cure', although a number of therapeutic interventions can modify the nystagmus waveform and/or reduce AHP. Nonetheless, very few interventions have thus far been shown to reliably improve visual function. In discussing the efficacy of any treatment for nystagmus, it is worth noting that VA cannot be relied upon as a sole outcome measure. Often, treatments that dampen nystagmus result in a *subjective* change in visual function, but no significant change using standard clinical measures such as VA. Since the foveas are only intermittently directed towards the point of interest, it has been suggested that a 'slowness to see' may be involved[8,9], which is not taken into account using VA testing alone. Therefore, it is possible that some treatments are more (or less) effective than previously reported. Treatment options can be divided into three broad groups:

- Optical
- Surgical
- Pharmacological

## Contact lenses

Contact lenses provide superior refractive correction over spectacles in nystagmus due to the reduction in peripheral lens aberrations and lack of induced prismatic effect that would be experienced as the eye moves away from the primary position with spectacles. Contact lenses are reported to provide at least a line of VA improvement beyond that afforded by spectacle wear[10,11]. Special care must be taken when fitting soft contact lenses in the presence of high astigmatism in nystagmus, as they may be more vulnerable to rotational instability with a consequential reduction in VA[12]. Hard contact lenses may be a more stable option in these cases[12]. Contact lenses also appear to dampen nystagmus eye movements in IN; an effect that is not present when the eye is anaesthetised[13]. This interesting finding suggests that the presence of the lens touching the eye – rather than the optical effects of the lens – serves to reduce nystagmus, implying that nystagmus intensity may be partially governed by signals from nerves in this region. Further work[14] has shown that cutaneous stimulation of the ophthalmic division of the trigeminal nerve can cause a reduction of nystagmus intensity, using gentle touches, vibrations, pressing and rubbing of the forehead and upper eyelids.

## Use and modification of the null zone

Using the *null zone* of gaze usually improves visual function in IN[15]. A specific head posture is used to achieve the required gaze angle. Whilst the use of such a head posture should not be dissuaded (to aid visual development), long-term use of an extreme head posture may result in a restricted range of movement of the neck[16]. Many of the therapeutic interventions available for IN aim to improve the availability of the null zone during general viewing.

Simple environmental adjustments can help when performing tasks for which a null zone is used. For example, Figure 1 demonstrates how a change in the seating position of a schoolchild with IN and an eccentric null zone may improve comfort for extended periods of viewing.

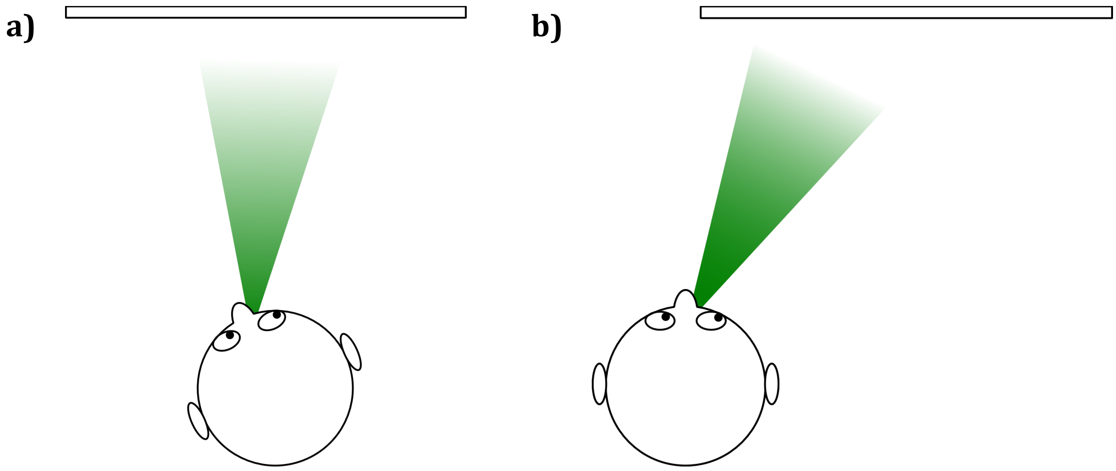


**Figure 1: Sitting a child with IN on the appropriate side of a classroom can make schoolwork more comfortable. (a): Child turns their head to place their null zone in the direction of the board. In (b), sitting the child on the left of the class allows them to maintain a comfortable head posture at school and still use the null zone.**

## Prism therapy

Prisms may be used to allow patients to adopt their null zone of gaze whilst keeping the head straight. For example, a patient with a null zone in rightgaze (head turn left) would benefit from base *left* prisms in front of each eye (Figure 2).

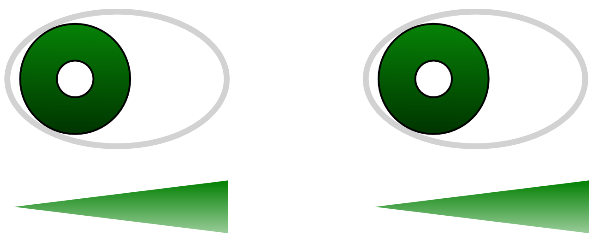

**Figure 2: Prismatic correction used to facilitate a rightward null zone (head turn left). Prisms are placed with the base toward the direction of the head turn (RE base *in*; LE base *out*).**

Many nystagmats have a convergent null zone[2,17]. Therefore, it is possible to dampen nystagmus using base out prisms (to induce convergence) . However, it is unclear whether the contrast sensitivity function can be improved in this way[17,18]. Rarely, the null zone is in the divergent position, in which case *base in* prism can reduce nystagmus intensity[19]. As always when prescribing prism, care must be taken to balance the refractive and prismatic prescriptions with respect to the *accommodative convergence : accommodation ratio*[18]. Accommodative convergence has been shown to have no effect on nystagmus, hence spherical manipulation of spectacle prescriptions does not improve nystagmus[17]. Typically, 7$^{\Delta}$ base out R&L are prescribed, with -1.00 DS to balance the prescription in pre-presbyopes[20].

## Surgical procedures

Surgical treatment for IN should be tailored to the underlying type of nystagmus and its specific associated features. There are three main indications for surgery in IN:

- Correct AHP
- Treat associated strabismus
- Improve visual function

Several surgical procedures have been advocated for IN. Those currently in use are described below.

### *Kestenbaum surgery*

In the early 1950s Anderson, Goto and Kestenbaum independently devised a surgical technique to move the null zone towards the primary gaze position, thus reducing any AHP present. Their rationale was to create a gaze palsy to the side of preferred fixation. Anderson proposed a recession of the horizontal yoke muscles acting as agonists in the direction of the head turn. Goto described a resection of the antagonistic muscle of each eye, whilst Kestenbaum advocated surgery on all four horizontal rectus muscles. Currently, augmented, modified, vertical and torsional versions of Kestenbaum surgery are being performed, which are reported to be effective at reducing AHPs to 10° or less in 50-100% of cases[21–43]. Improvements in VA (0-43% of cases) and recognition time (0.3 seconds) have also been reported following Kestenbaum surgery[44–47]. The presence of periodic alternating nystagmus (PAN) is a contraindication to Kestenbaum surgery as the AHP is likely to persist or change to the opposite direction post-operatively[48].

### *Artificial divergence*

In patients demonstrating convergence-induced nystagmus dampening, artificial divergence surgery can be used to induce an exodeviation which can be overcome by employing fusional convergence. This is achieved through bimedial rectus recessions, which can be titrated to the degree of fusional reserves available[49]. Artificial divergence surgery requires that patients are able to maintain motor fusion and have a measurable level of stereopsis.

A trial of prism correction should be performed as part of the pre-operative assessment in order to determine the degree of acceptability of artificial divergence surgery. Improvements in both AHP and VA (up to two Snellen lines) have been reported with artificial divergence surgery, which can also be combined with a Kestenbaum procedure[50–53].

### *Recession of all horizontal recti*

In cases without an AHP or in which PAN is present, large (12.0 mm) recessions of the four horizontal recti to the equator of the globe may be used. This procedure reduces nystagmus intensity by reducing the lever arm of the muscle action, with varying degrees of improvement in VA, AHP and recognition time reported in the literature[48,54–62].

### *Tenotomy and reattachment*

The *tenotomy* procedure, which involves severing the horizontal recti at the muscle insertion followed by reattachment *at the original site*, is thought to modify the proprioceptive loop in the extraocular muscles, resulting in broadening of the null zone and improvement in the nystagmus waveform and foveation duration. Although subjective improvements in VA have been reported, improvements in clinically measured VA appear to be limited (average 2.5 letters on an ETDRS chart)[63–66].

### *Combination surgery*

It is possible to perform combinations and modifications of the above surgical procedures whilst simultaneously correcting associated strabismus. These procedures are generally associated with positive outcomes, with improvements in nystagmus intensity and AHP, small improvements in VA (less than one line on a letter chart), and symptomatic relief of diplopia and oscillopsia reported[30,67–69].

## Pharmacological treatments

Many medicines are known to reduce the intensity of nystagmus. In 2002, a list of fourteen treatments reported to improve the condition was published[19]. Of these, four have been shown to significantly dampen nystagmus intensity in IN (memantine, gabapentin, cannabis and baclofen). Only memantine (20-40 mg daily) and gabapentin (up to 2400 mg daily) have been validated in a double-masked, randomised controlled trial, with both drugs demonstrating reductions in nystagmus intensity and improvements to VA (memantine, 0.15 logMAR; gabapentin, 0.09 logMAR) in comparison to placebo (0.04 logMAR)[70–72].

There have been several case reports demonstrating promising results from other medications. One case report of a subject with idiopathic IN showed that smoking cannabis reduced nystagmus intensity by 30% in the primary position of gaze, and improved clinical VA by 2-3 logMAR lines[73]. Baclofen is often used in patients with PAN, as it is known to reduce nystagmus amplitude, improve VA and alleviate AHP[74,75]. Dexedrine, a stimulant used to treat attention deficit hyperactivity disorder, has been shown to increase foveation duration, improve stereopsis, reduce exotropia magnitude, and improve VA in a patient with IN associated with rod-cone dystrophy[76]. More recently, it was demonstrated that brinzolamide, applied as topical eye drops, improved the nystagmus waveform in a patient with IN, providing hope for the development of non-systemic nystagmus pharmaceuticals[77].

The mechanisms of the above medications are currently unknown, although their mode of action is suspected to be through sedation rather than specifically reducing the eye movements[78]. For a detailed account of the pharmacological treatments available for IN, the reader is directed to recent review articles[79–81].

## Other therapies

A variety of therapies have been proposed in the past for treating IN. Although none have been shown to consistently improve measurable aspects of vision, there are several case reports of subjective visual improvements.

### *Biofeedback*

Auditory biofeedback, derived from live eye movement recordings, provide patients with the ability to ‘hear their nystagmus’, and with practice, patients can learn to consciously reduce nystagmus intensity by around 60%[57]. As with most IN treatments, the effect on visual function appears to be variable; one study reported changes between 0.13 and 0.32 logMAR[59], whereas one only reported a subjective awareness of improved vision[58]. Not all patients are able to apply the same techniques outside of the laboratory setting, but one individual was able to reduce their nystagmus to 50% of the pre-training level on demand, without biofeedback[60]. The effect is assumed to be similar to meditation, and as a result, studies into the effects of mindfulness meditation on IN are currently ongoing in the US.

### *Acupuncture*

Insertion of acupuncture needles into the sternocleidomastoid muscles of the neck may reduce nystagmus intensity in some individuals[61]. This can also improve the duration of foveation periods, with the effects sometimes being sustained after treatment[62].

## Management of acquired nystagmus

All of the optical and surgical options discussed above for the treatment of IN can be considered in suitable cases of acquired nystagmus. In addition, the treatment of acquired nystagmus can be customised to the underlying aetiology and type of nystagmus[80].

Acetazolamide and aminopyridines may be of benefit in acquired downbeat nystagmus, particularly in patients with a diagnosis of episodic ataxia type 2[82–85]. In cases of acquired downbeat nystagmus from a Chiari 1 malformation, a suboccipital craniectomy may be effective[86–88].

It is important to identify cases of acquired pendular nystagmus and PAN, as these forms may respond to pharmacological intervention. There is increasing evidence that the use of GABAergic agents such as gabapentin may suppress acquired pendular nystagmus[89–91]. Memantine, an NMDA receptor antagonist, is also effective in acquired pendular nystagmus[92]. Similar to PAN in IN, acquired PAN responds to baclofen treatment[19,93].

### Botox

Retrobulbar injections of clostridium botulinum A exotoxin (*Botox*) are occasionally used as a therapy for oscillopsia reduction in acquired nystagmus. The toxin causes a temporary extraocular muscle paralysis, with oscillopsia reduction lasting for 5-13 weeks[94]. Although oscillopsia is typically absent in IN, Botox injections have been shown to reduce nystagmus amplitude and improve VA in some individuals[95,96]. Despite low complication rates, due to the short duration of the effects, repeated injections are necessary to maintain therapeutic efficacy. In addition, paralysis of the extraocular muscles restricts patients' ability to execute normal eye movements.

### Magnetic implants

The use of magnetic implants in the orbit and globe have been proposed to anchor the eyeball at a specific gaze angle, thus reducing nystagmus. Trials are currently ongoing into this technology.

# Summary

The management options available for nystagmus depend on the type of nystagmus present, the underlying condition (if any), and nuances such as the position of the null zone (in IN) or specific sub-type (in acquired nystagmus). Seemingly small changes in clinical measures can have a significant impact on subjective visual function, so it is worthwhile for clinicians to have a working knowledge of the various therapeutic interventions available.

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