

Online Research @ Cardiff

This is an Open Access document downloaded from ORCA, Cardiff University's institutional repository: <http://orca.cf.ac.uk/93430/>

This is the author's version of a work that was submitted to / accepted for publication.

Citation for final published version:

Dunn, Matthew and Lee, Helena 2016. Management of nystagmus. *Optometry Today* 56 (8) , pp. 64-68. file

Publishers page:

Please note:

Changes made as a result of publishing processes such as copy-editing, formatting and page numbers may not be reflected in this version. For the definitive version of this publication, please refer to the published source. You are advised to consult the publisher's version if you wish to cite this paper.

This version is being made available in accordance with publisher policies. See <http://orca.cf.ac.uk/policies.html> for usage policies. Copyright and moral rights for publications made available in ORCA are retained by the copyright holders.



Management of nystagmus

Matt J. Dunn¹, Helena Lee²

¹ School of Optometry and Vision Sciences, Cardiff University, Cardiff, UK

² Southampton University Hospital, Southampton, UK

Correspondence to:

Matt J. Dunn, PhD, MCOptom, FHEA

Deputy Director, Research Unit for Nystagmus

School of Optometry and Vision Sciences

Cardiff University

Maindy Road

Cardiff CF24 4HQ, Wales, UK

Tel: +44 (0)29 20870551

Fax: +44 (0)29 20874859

Email: DunnMJ1@cardiff.ac.uk

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 world¹) 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 difficulties³. 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 nystagmus⁴. 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 VA⁵.

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 intensity⁶.

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 cases⁷.

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 VA¹². Hard contact lenses may be a more stable option in these cases¹². Contact lenses also appear to dampen nystagmus eye movements in IN; an effect that is not present when the eye is anaesthetised¹³. 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 work¹⁴ 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 IN¹⁵. 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 neck¹⁶. 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.

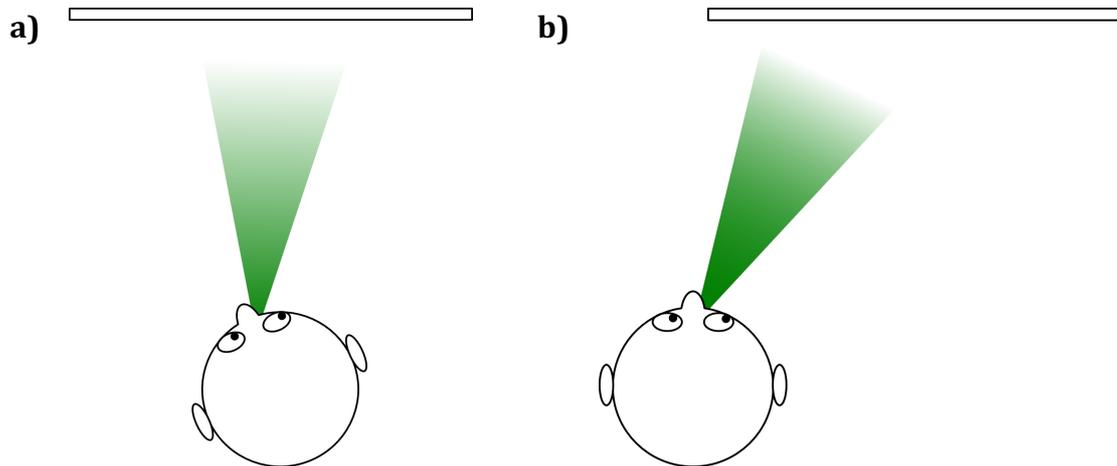


Figure 1: Sitting a child with IN on the appropriate side of a classroom can make schoolwork more comfortable. (a): Child turns their head to place their null zone in the direction of the board. In (b), sitting the child on the left of the class allows them to maintain a comfortable head posture at school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

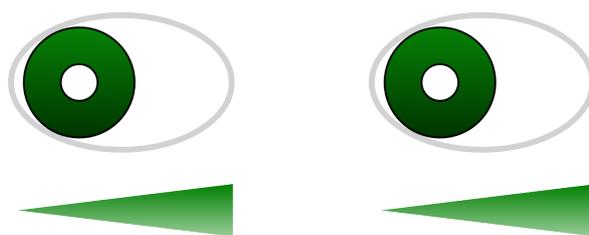


Figure 2: Prismatic correction used to facilitate a rightward null zone (head turn left). Prisms are placed with the base toward the direction of the head turn (RE base *in*; LE base *out*).

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 intensity¹⁹. 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 nystagmus¹⁷. Typically, 7^Δ base out R&L are prescribed, with -1.00 DS to balance the prescription in pre-presbyopes²⁰.

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 cases²¹⁻⁴³. Improvements in VA (0-43% of cases) and recognition time (0.3 seconds) have also been reported following Kestenbaum surgery⁴⁴⁻⁴⁷. 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-operatively⁴⁸.

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 available⁴⁹. 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 procedure⁵⁰⁻⁵³.

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 published¹⁹. 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 lines⁷³. 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 dystrophy⁷⁶. 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 pharmaceuticals⁷⁷.

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 movements⁷⁸. For a detailed account of the pharmacological treatments available for IN, the reader is directed to recent review articles⁷⁹⁻⁸¹.

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 logMAR⁵⁹, whereas one only reported a subjective awareness of improved vision⁵⁸. 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 biofeedback⁶⁰. 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 individuals⁶¹. This can also improve the duration of foveation periods, with the effects sometimes being sustained after treatment⁶².

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 nystagmus⁸⁰.

Acetazolamide and aminopyridines may be of benefit in acquired downbeat nystagmus, particularly in patients with a diagnosis of episodic ataxia type 2⁸²⁻⁸⁵. In cases of acquired downbeat nystagmus from a Chiari 1 malformation, a suboccipital craniectomy may be effective⁸⁶⁻⁸⁸.

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 nystagmus⁸⁹⁻⁹¹. Memantine, an NMDA receptor antagonist, is also effective in acquired pendular nystagmus⁹². 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 weeks⁹⁴. 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.

References

1. Brickner RM. Oscillopsia - A new symptom commonly occurring in multiple sclerosis. *Arch Neurol Psych.* 1936;36(3):586-589.
2. Abadi R V, Bjerre A. Motor and sensory characteristics of infantile nystagmus. *Br J Ophthalmol.* 2002;86(10):1152-1160.
3. McLean RJ, Windridge KC, Gottlob I. Living with nystagmus: a qualitative study. *Br J Ophthalmol.* 2012.
4. Sanders J. The UK Nystagmus Network (NN). *Semin Ophthalmol.* 2006;21:61.
5. Zubcov AA, Reinecke RD, Gottlob I, Manley DR, Calhoun JH. Treatment of manifest latent nystagmus. *Am J Ophthalmol.* 1990;110(2):160-7.
6. Calcutt C, Crook W. The treatment of amblyopia in patients with latent nystagmus. *Br Orthopt J.* 1972;29:70-72.
7. Sturm V, Hejcmanova M, Landau K. Effects of extraocular muscle surgery in children with monocular blindness and bilateral nystagmus. *BMC Ophthalmol.* 2014;14:137.
8. Dunn MJ, Margrain TH, Woodhouse JM, Erichsen JT. Visual processing in infantile nystagmus is not slow. *Invest Ophthalmol Vis Sci.* 2015;56(9):5094-5101.
9. Wang ZI, Dell'Osso LF. Being "slow to see" is a dynamic visual function consequence of infantile nystagmus syndrome: model predictions and patient data identify stimulus timing as its cause. *Vis Res.* 2007;47(11):1550-1560.
10. Allen ED, Davies PD. Role of contact lenses in the management of congenital nystagmus. *Br J Ophthalmol.* 1983;67(12):834-836.
11. Biousse V, Tusa RJ, Russell B, et al. The use of contact lenses to treat visually symptomatic congenital nystagmus. *J Neurol Neurosurg Psychiatry.* 2004;75(2):314-316.
12. Jayaramachandran P, Proudlock FA, Odedra N, Gottlob I, McLean RJ. A Randomized Controlled Trial Comparing Soft Contact Lens and Rigid Gas-Permeable Lens Wearing in Infantile Nystagmus. *Ophthalmology.* 2014.
13. Dell'Osso LF, Traccis S, Abel LA, Erzurum SI. Contact lenses and congenital nystagmus. *Clin Vis Sci.* 1988;3(3):229-232.
14. Dell'Osso LF, Leigh RJ, Daroff RB. Suppression of congenital nystagmus by cutaneous stimulation. *Neuro-Ophthalmology.* 1991;11(3):173-175.
15. Costa ACRV da, Lopes MCB, Nakanami CR. Influence of head posture on the visual acuity of children with nystagmus. *Arq Bras Oftalmol.* 2014;77(1):8-11.

16. Morris B, Smith V, Elphick J, Laws DE. Compensatory head posture and neck problems: is there an association? A cohort study of nystagmus patients. *Eye (Lond)*. 2009;23(2):279–83.
17. Dickinson CM. The elucidation and use of the effect of near fixation in congenital nystagmus. *Ophthalmic Physiol Opt*. 1986;6(3):303–311.
18. Dell’Osso LF. Development of new treatments for congenital nystagmus. *Ann N Y Acad Sci*. 2002;956:361–379.
19. Stahl JS, Plant GT, Leigh RJ. Medical treatment of nystagmus and its visual consequences. *J R Soc Med*. 2002;95(5):235–237.
20. Khanna S, Dell’Osso LF. The diagnosis and treatment of infantile nystagmus syndrome (INS). *ScientificWorldJournal*. 2006;6:1385–1397.
21. Calhoun JH, Harley RD. Surgery for abnormal head position in congenital nystagmus. *Trans Am Ophthalmol Soc*. 1973;71:70–83; discussion 84–7.
22. Sandall GS. Surgical treatment of congenital nystagmus in patients with singular binocular vision. *Ann Ophthalmol*. 1976;8(2):227–38.
23. Nelson LB, Ervin-Mulvey LD, Calhoun JH, Harley RD, Keisler MS. Surgical management for abnormal head position in nystagmus: the augmented modified Kestenbaum procedure. *Br J Ophthalmol*. 1984;68(11):796–800.
24. Scott WE, Kraft SP. Surgical treatment of compensatory head position in congenital nystagmus. *J Pediatr Ophthalmol Strabismus*. 1984;21(3):85–95.
25. Pratt-Johnson JA. Results of surgery to modify the null-zone position in congenital nystagmus. *Can J Ophthalmol*. 1991;26(4):219–23.
26. Kraft SP, O’Donoghue EP, Roarty JD. Improvement of compensatory head postures after strabismus surgery. *Ophthalmology*. 1992;99(8):1301–8.
27. Lee IS, Lee JB, Kim HS, Lew H, Han SH. Modified Kestenbaum surgery for correction of abnormal head posture in infantile nystagmus: outcome in 63 patients with graded augmentaton. *Binocul Vis Strabismus Q*. 2000;15(1):53–8.
28. Chang Y-H, Chang JH, Han S-H, Lee JB. Outcome study of two standard and graduated augmented modified Kestenbaum surgery protocols for abnormal head postures in infantile nystagmus. *Binocul Vis Strabismus Q*. 2007;22(4):235–41.
29. Kang NY, Isenberg SJ. Kestenbaum procedure with posterior fixation suture for anomalous head posture in infantile nystagmus. *Graefe’s Arch Clin Exp Ophthalmol = Albr von Graefes Arch für Klin und Exp Ophthalmol*. 2009;247(7):981–7.
30. Kumar A, Shetty S, Vijayalakshmi P, Hertle RW. Improvement in visual acuity following surgery for correction of head posture in infantile nystagmus syndrome. *J Pediatr Ophthalmol Strabismus*. 2011;48(6):341–346.
31. Wang P, Lou L, Song L. Design and efficacy of surgery for horizontal idiopathic nystagmus with abnormal head posture and strabismus. *J Huazhong Univ Sci*

- Technolog Med Sci.* 2011;31(5):678–81.
32. Schild AM, Thoenes J, Fricke J, Neugebauer A. Kestenbaum procedure with combined muscle resection and tucking for nystagmus-related head turn. *Graefes Arch Clin Exp Ophthalmol.* 2013.
 33. Bérard P V, Spielmann A, Reydy R. [Surgery using Cuppers' wire. Physiological bases. Technics and indications]. *Bull des sociétés d'ophtalmologie Fr.* 1976;76(12):1111–6.
 34. Pierse D. Operation on the vertical muscles in cases of nystagmus. *Br J Ophthalmol.* 1959;43(4):230–3.
 35. Parks MM. Symposium: nystagmus. Congenital nystagmus surgery. *Am Orthopt J.* 1973;23:35–9.
 36. Roberts EL, Saunders RA, Wilson ME. Surgery for vertical head position in null point nystagmus. *J Pediatr Ophthalmol Strabismus.* 1996;33(4):219–24.
 37. Hertle RW, Yang D, Adams K, Caterino R. Surgery for the treatment of vertical head posturing associated with infantile nystagmus syndrome: results in 24 patients. *Clin Exp Ophthalmol.* 2011;39(1):37–46.
 38. Yang MB, Pou-Vendrell CR, Archer SM, Martonyi EJ, Del Monte MA. Vertical rectus muscle surgery for nystagmus patients with vertical abnormal head posture. *J AAPOS.* 2004;8(4):299–309.
 39. Lee J. Surgical management of nystagmus. *J R Soc Med.* 2002;95(5):238–241.
 40. Lueder GT, Galli M. Oblique muscle surgery for treatment of nystagmus with head tilt. *J AAPOS.* 2012;16(4):322–326.
 41. Conrad HG, DeDecker W. Torsional Kestenbaum procedure evolution of a surgical concept. In: Reinecke RD, ed. *Proceedings of the Fourth Meeting of the International Strabismological Association.* Orlando: Grune and Stratton; 1982:301–314.
 42. Spielmann A. [Oblique Kestenbaum's technic on the rectus muscles. Personal technic]. *Bull des sociétés d'ophtalmologie Fr.* 1987;87(7-8):919–24.
 43. von Noorden GK, Jenkins RH, Rosenbaum AL. Horizontal transposition of the vertical rectus muscles for treatment of ocular torticollis. *J Pediatr Ophthalmol Strabismus.* 1993;30(1):8–14.
 44. Flynn JT, Dell'Osso LF. The effects of congenital nystagmus surgery. *Ophthalmology.* 1979;86(8):1414–27.
 45. Ugurbas SC, Baker JD. Secondary or new compensatory head posture after Anderson-Kestenbaum surgery. *Eur J Ophthalmol.* 22(2):131–5.
 46. ElKamshoushy A, Shawky D, Elmassry A, Elbaha S, Abdel Wahab MM, Sprunger D. Improved visual acuity and recognition time in nystagmus patients following four-muscle recession or Kestenbaum-Anderson procedures. *J AAPOS.*

- 2012;16(1):36–40.
47. Abadi R V, Whittle J. Surgery and compensatory head postures in congenital nystagmus. A longitudinal study. *Arch Ophthalmol (Chicago, Ill 1960)*. 1992;110(5):632–5.
 48. Gradstein L, Reinecke RD, Wizov SS, Goldstein HP. Congenital periodic alternating nystagmus. Diagnosis and management. *Ophthalmology*. 1997;104(6):928–929.
 49. Spielmann A. Clinical rationale for manifest congenital nystagmus surgery. *J AAPOS*. 2000;4(2):67–74.
 50. Sendler S, Shallo-Hoffmann J, Mühlendyck H. [Artificial divergence surgery in congenital nystagmus]. *Fortschr Ophthalmol*. 1990;87(1):85–9.
 51. Cüppers C. [Problems in the surgery for ocular nystagmus]. *Klin Monatsblätter für Augenheilkd*. 1971;159(2):145–57.
 52. Zubcov AA, Stark N, Weber A, Wizov SS, Reinecke RD. Improvement of visual acuity after surgery for nystagmus. *Ophthalmology*. 1993;100(10):1488–1497.
 53. Gräf M, Droutsas K, Kaufmann H. Surgery for nystagmus related head turn: Kestenbaum procedure and artificial divergence. *Graefe's Arch Clin Exp Ophthalmol = Albr von Graefes Arch für Klin und Exp Ophthalmol*. 2001;239(5):334–41.
 54. Atilla H, Erkam N, Işıkçelik Y. Surgical treatment in nystagmus. *Eye*. 1999;13(1):11–15.
 55. Helveston EM, Ellis FD, Plager DA. Large recession of the horizontal recti for treatment of nystagmus. *Ophthalmology*. 1991;98(8):1302–1305.
 56. Alió JL, Chipont E, Mulet E, De La Hoz F. Visual performance after congenital nystagmus surgery using extended hang back recession of the four horizontal rectus muscles. *Eur J Ophthalmol*. 2003;13(5):415–423.
 57. Boyle NJ, Dawson ELM, Lee JP. Benefits of retroequatorial four horizontal muscle recession surgery in congenital idiopathic nystagmus in adults. *J AAPOS*. 2006;10(5):404–408.
 58. Hertle RW, Dell'Osso LF. Benefits of retroequatorial four horizontal muscle recession surgery in congenital idiopathic nystagmus in adults. *J AAPOS*. 2007;11(3):313.
 59. Davis PL, Baker RS, Piccione RJ. Large recession nystagmus surgery in albinos: effect on acuity. *J Pediatr Ophthalmol Strabismus*. 34(5):279–83; discussion 283–5.
 60. Sprunger DT, Fahad B, Helveston EM. Recognition time after four muscle recession for nystagmus. *Am Orthopt J*. 1997;47:122–125.
 61. von Noorden GK, Sprunger DT. Large rectus muscle recessions for the treatment of congenital nystagmus. *Arch Ophthalmol (Chicago, Ill 1960)*. 1991;109(2):221–

- 4.
62. Bagheri A, Farahi A, Yazdani S. The effect of bilateral horizontal rectus recession on visual acuity, ocular deviation or head posture in patients with nystagmus. *J AAPOS*. 2005;9(5):433–7.
 63. Dell’Osso LF. Extraocular muscle tenotomy, dissection, and suture: a hypothetical therapy for congenital nystagmus. *J Pediatr Ophthalmol Strabismus*. 1998;35(4):232–233.
 64. Dell’Osso LF, Hertle RW, Williams RW, Jacobs JB. A new surgery for congenital nystagmus: effects of tenotomy on an achiasmatic canine and the role of extraocular proprioception. *J AAPOS*. 1999;3(3):166–182.
 65. Hertle RW, Dell’Osso LF, FitzGibbon EJ, Thompson D, Yang D, Mellow SD. Horizontal rectus tenotomy in patients with congenital nystagmus: results in 10 adults. *Ophthalmology*. 2003;110(11):2097–2105.
 66. Wang ZI, Dell’osso LF, Prakash S, Chen X. Smooth-pursuit changes after the tenotomy and reattachment procedure for infantile nystagmus syndrome: model predictions and patient data. *J Pediatr Ophthalmol Strabismus*. 2012;49(5):295–302.
 67. Bagheri A, Aletaha M, Abrishami M. The effect of horizontal rectus muscle surgery on clinical and eye movement recording indices in infantile nystagmus syndrome. *Strabismus*. 2010;18(2):58–64.
 68. Wang ZI, Dell’Osso LF, Tomsak RL, Jacobs JB. Combining recessions (nystagmus and strabismus) with tenotomy improved visual function and decreased oscillopsia and diplopia in acquired downbeat nystagmus and in horizontal infantile nystagmus syndrome. *J AAPOS*. 2007;11(2):135–141.
 69. Bishop JE. A novel new [yet again] procedure for correction of compensatory head posture in infantile nystagmus; augmented Anderson plus Dell’Osso-Hertle. *Binocul Vis Strab Q Simms Rom*. 2011;26(1):37–42.
 70. Sarvananthan N, Proudlock FA, Choudhuri I, Dua H, Gottlob I. Pharmacologic treatment of congenital nystagmus. *Arch Ophthalmol*. 2006;124(6):916–918.
 71. Shery T, Proudlock FA, Sarvananthan N, McLean RJ, Gottlob I. The effects of gabapentin and memantine in acquired and congenital nystagmus: a retrospective study. *Br J Ophthalmol*. 2006;90(7):839–843.
 72. McLean RJ, Proudlock F, Thomas S, Degg C, Gottlob I. Congenital nystagmus: randomized, controlled, double-masked trial of memantine/gabapentin. *Ann Neurol*. 2007;61(2):130–138.
 73. Pradeep A, Thomas S, Roberts EO, Proudlock FA, Gottlob I. Reduction of congenital nystagmus in a patient after smoking cannabis. *Strabismus*. 2008;16(1):29–32.
 74. Solomon D, Shepard N, Mishra A. Congenital periodic alternating nystagmus: response to baclofen. *Ann N Y Acad Sci*. 2002;956:611–615.

75. Comer RM, Dawson EL, Lee JP. Baclofen for patients with congenital periodic alternating nystagmus. *Strabismus*. 2006;14(4):205–209.
76. Hertle RW, Maybodi M, Bauer RM, Walker K. Clinical and oculographic response to Dexedrine in a patient with rod-cone dystrophy, exotropia, and congenital aperiodic alternating nystagmus. *Binocul Vis Strabismus Q*. 2001;16(4):259–264.
77. Dell’Osso LF, Hertle RW, Leigh RJ, Jacobs JB, King S, Yaniglos S. Effects of topical brinzolamide on infantile nystagmus syndrome waveforms: eyedrops for nystagmus. *J Neuroophthalmol*. 2011;31(3):228–233.
78. Abel LA. Infantile nystagmus: current concepts in diagnosis and management. *Clin Exp Optom*. 2006;89(2):57–65.
79. McLean RJ, Gottlob I. The pharmacological treatment of nystagmus: a review. *Expert Opin Pharmacother*. 2009;10(11):1805–1816.
80. Strupp M, Thurtell MJ, Shaikh AG, Brandt T, Zee DS, Leigh RJ. Pharmacotherapy of vestibular and ocular motor disorders, including nystagmus. *J Neurol*. 2011;258(7):1207–1222.
81. Thurtell MJ, Leigh RJ. Treatment of nystagmus. *Curr Treat Options Neurol*. 2012;14(1):60–72.
82. Ilg W, Bastian AJ, Boesch S, et al. Consensus paper: management of degenerative cerebellar disorders. *Cerebellum*. 2014;13(2):248–68.
83. Riant F, Vahedi K, Tournier-Lasserre E. [Hereditary episodic ataxia]. *Rev Neurol (Paris)*. 2011;167(5):401–7.
84. Kalla R, Spiegel R, Claassen J, et al. Comparison of 10-mg doses of 4-aminopyridine and 3,4-diaminopyridine for the treatment of downbeat nystagmus. *J Neuroophthalmol*. 2011;31(4):320–5.
85. Strupp M, Brandt T. Current treatment of vestibular, ocular motor disorders and nystagmus. *Ther Adv Neurol Disord*. 2009;2(4):223–39.
86. Goodwin D, Halvorson AR. Chiari I malformation presenting as downbeat nystagmus: clinical presentation, diagnosis, and management. *Optometry*. 2012;83(2):80–6.
87. Spooner JW, Baloh RW. Arnold-Chiari malformation: improvement in eye movements after surgical treatment. *Brain*. 1981;104(Pt 1):51–60.
88. Pedersen RA, Troost BT, Abel LA, Zorub D. Intermittent downbeat nystagmus and oscillopsia reversed by suboccipital craniectomy. *Neurology*. 1980;30(11):1239–42.
89. Averbuch-Heller L, Tusa RJ, Fuhry L, et al. A double-blind controlled study of gabapentin and baclofen as treatment for acquired nystagmus. *Ann Neurol*. 1997;41(6):818–25.
90. Stahl JS, Rottach KG, Averbuch-Heller L, von Maydell RD, Collins SD, Leigh RJ. A

- pilot study of gabapentin as treatment for acquired nystagmus. *Neuroophthalmology*. 1996;16(2):107–13.
91. Fabre K, Smet-Dieleman H, Zeyen T. Improvement of acquired pendular nystagmus by gabapentin: case report. *Bull la Société belge d'ophtalmologie*. 2001;(282):43–6.
 92. Starck M, Albrecht H, Pöllmann W, Straube A, Dieterich M. Drug therapy for acquired pendular nystagmus in multiple sclerosis. *J Neurol*. 1997;244(1):9–16.
 93. Straube A, Leigh RJ, Bronstein A, et al. EFNS task force--therapy of nystagmus and oscillopsia. *Eur J Neurol*. 2004;11(2):83–9.
 94. Helveston EM, Pogrebniak AE. Treatment of acquired nystagmus with botulinum A toxin. *Am J Ophthalmol*. 1988;106(5):584–586.
 95. Carruthers J. The treatment of congenital nystagmus with Botox. *J Pediatr Ophthalmol Strabismus*. 1995;32(5):306–308.
 96. Hernández-García E, Gómez-De-Liaño-Sánchez R. Use of botulinum toxin in a patient with pendular congenital nystagmus. *Arch Soc Esp Ophthalmol*. 2012;87(10):330–332.