

Article ▶ A Life with Nystagmus

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ABSTRACT

Nystagmus is one of the major concerns of visual impairment globally affecting quality of life. It can severely disrupt the lifestyle of those with nystagmus, especially deteriorating confidence and self esteem. There is no clear understanding about the impact of nystagmus on social and visual functions. The effect of nystagmus on driving performance can be severe, and very few with this condition can drive. This directly or indirectly affects their daily living and hampers routine activities, leading to the use of public transportation for routine activities. Current medical treatment has not focused upon improving the quality of living and development of effective therapies. This qualitative review elaborates critically on the quality of living with nystagmus, driving difficulties, and the current treatment options available. The review also emphasizes the pressing need for focused research on various aspects of nystagmus.

Keywords: nystagmus, null point, oscillations, oscillopsia, quality of life, vision therapy

Nystagmus is an involuntary eye movement in a 'to and fro' manner and presents with a rhythmic, pendular, or jerky conjugate oscillation of the eyes. It can be infantile or acquired later in life. Infantile Nystagmus (INS) is further classified as: 1) pure infantile nystagmus syndrome, 2) spasmus nutans syndrome; and 3) nystagmus associated with ocular disease.¹ Acquired nystagmus can be present in neurologic and vestibular diseases. Vestibular nystagmus is caused by inner ear semi-circular canal dysfunction. Acquired nystagmus is also likely to result from abnormal development or pathologic malfunction of areas in the cognitive system and gaze stability or afferent pathway disorders.² The understanding of pathologic mechanisms in nystagmus is quite anecdotal but has increased recently. Mutation in a novel gene (FRMD7) has been found in association with pure X-linked infantile nystagmus.³ The amplitude and frequency of the nystagmus indicates the severity of visual function loss. The eyes and/or the head move sideways, and the person may cock their head to view through a position where the frequency of nystagmus is least.

A common myth is that people with nystagmus see the stationary objects moving in conjunction with their eye movement; however, this is not exactly true. They can see the objects similarly to an individual without a visual impairment but present with reduced visual acuity and difficulty maintaining fixation while concentrating on an object. An individual with congenital motor nystagmus can achieve a null point by moving the eyes or head to a particular gaze. Inappropriate saccades, including intrusions and oscillations, may hamper vision⁴ and are one of the causes of reduced vision.

Prevalence of Nystagmus

The prevalence of nystagmus in the general population is not clearly defined as very few evaluative studies have been performed. A cohort study conducted on partially sighted or blind children older than 15 years in Denmark estimated the prevalence of nystagmus.⁵ In the Netherlands, a study was

conducted among 220,802 army recruits, who were excluded from service because of deteriorated vision.⁶ It was reported that 117 were discharged because of nystagmus. Out of those discharged, 56 were examined further, and 29 were diagnosed with congenital type nystagmus. Another study evaluated elementary school children in first grade in Malmö, Sweden between 1941 and 1959, which was further extended and done on the family members of affected children.⁷ This study reported a total frequency of 1 in 1500 children; boys: 1 in 1000 and girls: 1 in 2800. This study examined only children with nystagmus and their affected family members but excluded adults with non-familial forms of nystagmus. In the United Kingdom (UK), a study was performed on 10-year old children in a representative sample of 15,000 with visual acuities ranging from 20/20 to poorer than 20/200.⁸ Congenital nystagmus was reported as the second most common cause of partial sight or blindness following congenital cataract. However, none of these studies provided data on adults or children with acquired nystagmus. A recent study reports the prevalence of nystagmus in the general population to be 24.0 per 10,000 population.⁹ This study reported that the most common form of nystagmus was neurological in origin, which accounted for 6.8 per 10,000 population. In addition, nystagmus associated with low vision was reported to be 4.2 per 10,000 population, and nystagmus with retinal disease association was reported to be 3.4 per 10,000 population.

Quality of Life with Nystagmus

Nystagmus can severely disrupt individuals' lifestyles, deteriorating confidence and self-esteem. The impact of nystagmus on visual (ability to perform activities of daily living) and social function (self perception of well-being, interaction with friends, relatives, colleagues, and contribution to society) is unclear. The literature available on the subject is vague and needs to be enhanced through greater research efforts. Not a single study has investigated visual function and quality of

life in Indian nystagmus patients. A study in the UK indicates that nystagmus is associated with very low visual function, and a strong correlation was found to be present between visual and social impairment.¹⁰ This study also assessed the parents and their ability to judge the impact of nystagmus on their children. It was found that the parents underestimate the impact of nystagmus.

Children with nystagmus were found to have lower social function than adults in the areas of appearance and confidence. Adults with nystagmus have been found to be at lower levels in career and education. There are multiple studies which report that the lifestyle of a child can severely affect his well-being, development, and adult health.¹¹⁻¹³ Studies conducted on nystagmoid patients with strabismus and/or amblyopia to assess the quality of life indicate interrupted schooling, lower job choice, and social activity.^{11,14} An individual's experience with difficulties in daily activities is determined jointly by a combination of visual, functional, psychological, social, and economic factors.¹⁵ The National Eye Institute Visual Function Questionnaire (NEI-VFQ),¹⁶ which is specifically designed to assess vision, is sensitive enough to determine functional status secondary to visual impairment. Vision-related quality of life (QOL) tools (disease-specific rather than generic tools) are able to detect the impact of a particular eye condition on an individual.^{17,18}

A recent study on the quality of life with nystagmus states six specific domains which are adversely affected: visual function, restriction of movement, standing out/not fitting in, feelings about the inner self, negativity about the future, and relationships.¹⁹ The cosmetic effect of nystagmus has not been studied properly; however, it has very similar characteristics as that of strabismus in which there is low self-esteem, affected interpersonal relationships, and lower self-image.²⁰ Hence, the overall quality of life is severely affected due to cosmetic appearance.

Driving with Nystagmus

Driving is a highly rigorous task in which vision plays a pivotal role.²¹ Various factors like visual acuity, contrast sensitivity, colour vision, and depth perception contribute to better visual performance while driving. Visual fields are considered superior to other visual functions while driving.²² A standardization in visual requirements for driving is highly essential for the safety of the driver as well as the people on the roads. Few studies have emphasized the need for visual field standards²² and the medical review requirements for driving;²³ however, the standards vary from one country to another. Casson et al.²² concluded that adequate contrast sensitivity is as important as good visual acuity for driving and emphasized the need for focused researches to determine the relation between the location of the visual field defect and fitness to drive. Owsley et al.²³ focused on valid and reliable assessment tools for the safety of low-vision drivers who use bioptic telescopes, the impact of monocular vision impairment on safety, and

the effectiveness of vision rescreening policies after initial licensure. The standard for visual need comes from evidence-based study; however, there is no focused study to address the visual standards and need in nystagmus. There is a pressing need for studies to assess the driving difficulties faced by people with nystagmus and the optimal visual requirements for such patients. The impact of nystagmus on driving performance can be severe, and very few with this condition drive. This directly or indirectly affects their daily living and hampers routine activities. Persons with nystagmus either have to use the public transit system or need to be dependent upon somebody for routine transportation because of their inability to drive and poor handling of the vehicle.

India has one of the largest highway and road networks, second only to the road network of the United States. Statistics indicate that over 140,000 people are killed and 510,000 people sustain injuries every year on Indian roads, and the data may still go much higher (>60% rise) by 2020.²⁴ The data indicates that either the visual standard for driver's licensing is not followed in India or there is a lack of a proper system to monitor drivers with poor vision involved in accidents. Prasad et al.²⁵ reported 40 truck drivers (age group 41-60 years) with defective vision out of 140 screened truck drivers and suggested that the drivers of this age group should be screened frequently to detect visual defects. People who are visually impaired and with nystagmus can have life-threatening accidents on highways and busy roads. Vision is an important aspect of driving, and improving the standards could potentially reduce road accidents. Hence, the assessment of vision and driving performance of people with nystagmus is of utmost priority. Reports from the UK indicate that in the year 1992, road accidents led to huge financial costs because of 30,000 injuries and 5000 fatalities. The economic cost to the UK in that year was estimated at £5.5 billion, and only a 10% reduction in these accidents could save £0.55 billion.

There is no clear evidence to link the road accidents to poor vision as vision data pertaining to drivers involved in accidents is not recorded properly. Hence, the registered blind or persons with substandard visual acuity should not be allowed to drive, and vision should be checked precisely for licensing. The vision standards in the UK are based upon the number plate eye test, where a person needs to read the number plate fixed to a motor vehicle at a certain distance.^{26,27} The current vision requirement for group 1 drivers in the UK is: visual acuity 6/12 with both eyes together for distance with corrective lenses if needed, peripheral vision 120 degrees horizontal, 20 degrees up and down, and no visual field loss in the central 20 degrees.²⁶ A person with nystagmus needs to report to the Driving and Vehicle Licensing Agency (DVLA) about the medical condition; failing to do so may lead to a fine up to £1,000 or prosecution. There are no specific guidelines or vision standards for visually impaired people in India.

A recent study indicates that inability to drive with nystagmus imposes severe restrictions in travel and at the same

Table 1. Treatment Options for Nystagmus

| Treatment approach | Examples |
|------------------------------|---|
| Medical | Gabapentin, Memantine, 4-aminopyridine, 3,4-diaminopyridine, Baclofen, Clonazepam, Valproate, Trihexyphenidyl, Benztropine, Scopolamine, Isoniazid, Carbamazepine, Barbiturates, Alcohol, Acetazolamide, Brinzolamide (topical), Cannabis |
| Optical | Contact lenses Contact lens and spectacle combinations Prisms Electro-optical devices |
| Surgical | Anderson-Kestenbaum procedure Cüppers' divergence procedure Recession of rectus muscles Tenotomy and reattachment procedure |
| Other (Miscellaneous) | Botulinum toxin Acupuncture Biofeedback Cutaneous stimulation |

time affects occupation.¹⁹ Most of the participants in the study reported use of public transportation. The evidence remains controversial and anecdotal concerning visual performance in nystagmus.

Current Treatment and Rehabilitation Measures for Nystagmus

Medical treatment primarily focuses on influencing the neurotransmitters with oral medication, such as baclofen, clonazepam, valproic acid, and gabapentin. The usefulness of these medications has been limited because of side effects; hence, the success of these drugs varies. Use of topical β -blockers such as betaxolol has been reported in a case of superior oblique myokymia.²⁸ Botulinum toxin is employed in dampening the amplitude, which is particularly useful in acquired forms of nystagmus accompanied by disturbing oscillopsia. However, the effect of botulinum toxin is not permanent, and injections have to be given repeatedly every 4 to 20 weeks. Helveston and Pogrebniak²⁹ have shown an improvement in visual acuity, from 20/80 to 20/30, in one patient with severe oscillopsia using a retrobulbar method. Occluding contact lenses and acupuncture are also effective therapies for nystagmus. Tinted occluding contact lenses have been proven to be successful in patients with acquired rotational nystagmus in both eyes causing dizziness and nausea due to multiple moving objects.³⁰ Non-opaque lenses have been previously used to treat congenital nystagmus.³¹ Acupuncture has been reported to be an effective treatment option for nystagmus. Improved foveation has been reported in four of six patients when the two needles were inserted into each sternocleidomastoid muscle belly.³² Vibratory and electrical stimulation have also shown to be effective in increasing the foveation; however, it is unclear whether this is secondary to the cervical afferents

or because of placebo effect.³² Vision therapy³³⁻³⁶ has also demonstrated improved visual function in individuals with nystagmus, though no studies have been done.

There is a paucity of evidence regarding the therapies for nystagmus. There is a lack of control groups in the studies focused on therapies; hence, it is difficult to differentiate between therapeutic and placebo effects. After-image feedback and intermittent photopic stimulation have been reported to be effective therapies.³⁷ Recent studies indicate that carbonic anhydrase inhibitors can suppress infantile nystagmus, including oral acetazolamide and topical brinzolamide.^{38,39} Infantile nystagmus can also be dampened after smoking cannabis.⁴⁰ Gene therapy is the potential hope for treatment of nystagmus associated with congenital retinal disorders. Studies reported that in an animal model of Leber's congenital amaurosis, successful gene therapy restored vision and reduced the associated nystagmus.⁴¹⁻⁴⁴ Thurtell⁴⁵ has proposed a few treatment approaches for nystagmus, which are summarized in Table 1.

Conclusion

We present for the first time a major qualitative review on the quality of life with nystagmus. Nystagmus can severely disrupt day-to-day activities with deterioration in self-esteem and confidence. Driving is one of the major areas affected, which forces the person to be dependent upon others for daily transportation and activities. There is a pressing need for focused research on the quality of living with nystagmus. Nystagmus is a global concern of visual impairment and reduced visual performance.

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