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NYSTAGMUS IN CHILDHOOD: A SYSTEMATIC REVIEW

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Abstract

Nystagmus is an involuntary rhythmic oscillation of the eyes that results in decreased visual acuity owing to the excessive motion of pictures on the retina. This is because nystagmus causes the eyes to move in a repetitive pattern. It is of the utmost importance to differentiate infantile nystagmus from acquired nystagmus. In order to accomplish this, it is necessary to take into consideration not only the time at which the nystagmus first appeared, but also the features of its waveform. Oscillations of the eyes that are involuntary, often conjugate, and frequently rhythmic are referred to as nystagmus. Infantile nystagmus syndrome (INS) is the condition that accounts for the majority of cases of nystagmus in children. Because this ailment can last a person's entire life, it is essential that they are familiar with all of the treatment choices that are at their disposal. The primary foci of this review are the underlying etiology of nystagmus, the psychosocial and functional implications of nystagmus, and the current methods of treatments, which include optical, pharmacological, surgical, and rehabilitative treatment approaches. At this time, the brain mechanisms that are responsible for INS are not completely known. The evidence is limited to mostly pre- and post-study designs, and there are few objective comparisons of treatment procedures. The treatment choices are meant to increase the foveation length or correct abnormal head positions. Individualized management of INS should be implemented. The patient and the patient's physician are the ones who should make the decision regarding which treatment will be most beneficial for the patient in question.

Kata kunci: *Childhood; Eyes; Nystagmus; Ophtalmology*

INTRODUCTION

Nystagmus is an involuntary rhythmic oscillation of the eyes, which leads to reduced visual acuity due to the excessive motion of images on the retina. It is essential to distinguish infantile from acquired nystagmus. This can be accomplished by taking into account not only the time of onset of the nystagmus, but also its waveform characteristics.[1,2] When the nystagmus is asymmetrical or unilateral, neurological illness should be suspected. In order to rule out any underlying ocular or systemic pathology in a child who has nystagmus, a thorough work-up that includes electrophysiology, laboratory tests, neurological examinations, and imaging may be required.[3]

Infantile nystagmus (IN) typically manifests in the first three to six months of life, whereas acquired nystagmus (AN) manifests later in life. IN can be idiopathic or associated with albinism, retinal disease, limited vision, or visual deprivation in early life, such as congenital cataracts, optic nerve hypoplasia, and retinal dystrophies, or it can be a component of neurological syndromes and neurologic diseases. It is possible to classify it as either Infantile Nystagmus Syndrome (INS) or acquired nystagmus, depending on the severity.[4,5]

It is estimated that there are 24 people in every 10,000 who have nystagmus in the UK.[5] In most cases, the initial symptoms of INS appear within the first six months of a person's life (the average age of onset is 1.9 months). Depending on the underlying condition, children who experience nystagmus may have a visual acuity (VA) that is virtually normal or they may have a visual acuity that is substantially compromised. Nystagmus, in any form, can be associated with severe visual loss; nevertheless, VA is not a comprehensive assessment of visual function.[6,7]

Infantile nystagmus prevalence rates have been reported largely from Europe and Asia, and they range from 1/1000 to 1/500,000. The lower the number, the less common the condition is. The most current study to provide prevalence data on nystagmus in North America comes from a small study that was conducted in which roughly 2,500 children were checked for ocular abnormalities, and 1 out of 283 had nystagmus. The study was conducted in Canada. There is also a lack of reliable data regarding the incidence of nystagmus.[8,9] This article provided about nystagmus in childhood.

METHODS

This systematic review followed the Preferred Reporting Items for Systematic Review and Meta-Analysis (PRISMA) 2020 criteria for its methodology. This systematic review was conducted to evaluate adolescent nystagmus. Examined subject matter is the focus of the studies under consideration. To effectively evaluate existing studies, it is essential that they meet certain criteria, including: 1) Articles must be available online for easy access; 2) Articles written in English are preferred; and 3) The systematic review will only consider articles published between 2015 and the present.

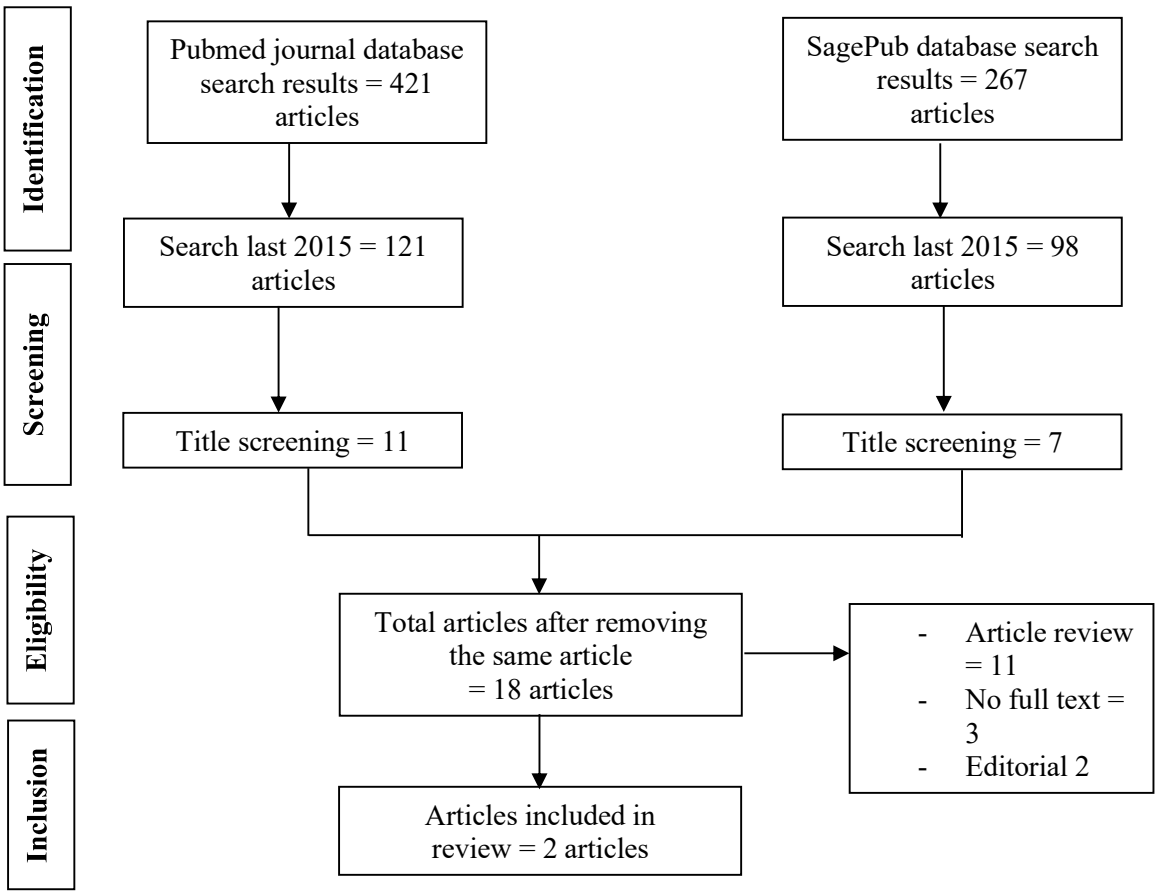

Figure 1. Article search flowchart

The search for studies to be included in the systematic review was carried out from May 12th, 2023 using the PubMed and SagePub databases by inputting the words: "nystagmus" and "childhood". Where *("nystagmus, pathologic"[MeSH Terms] OR ("nystagmus"[All Fields] AND "pathologic"[All Fields]) OR "pathologic nystagmus"[All Fields] OR "nystagmus"[All Fields]) AND ("childhood"[All Fields] OR "childhoods"[All Fields])* is used as search keywords.

The inclusion and exclusion criteria for the study were revised after a comprehensive literature review based on an examination of the titles and abstracts of previously published research. In the systematic review, only research initiatives that met all of the requirements were included. When comparing one research study to another, it is essential to consider the study's title, author, publication date, country of origin, research design, and variables being studied.

This content has been presented in a particular format for your consideration and evaluation. The authors conducted independent evaluations of a selection of research endeavors described in the titles and abstracts of the publications to determine if the studies were admissible. Then, the full texts of the studies that satisfy the inclusion criteria for the systematic review will be evaluated to determine which publications qualify for categorical inclusion in the review.

RESULT

Nash, et al (2017)[4] study showed seventy-one children were diagnosed during the 30-year period, resulting in an annual incidence of 6.72 per 100,000 children younger than 19 years (95% confidence interval [CI] = 5.15–8.2). Infantile nystagmus with onset by 6 months was present in 62 (87.3%) of the study patients, equating to a birth prevalence of 1 in 821. The median age of the cohort at diagnosis was 12.7 months (range: 0 days to 18.6 years), and 42 (59.2%) were males. In descending order.

The most prevalent types of nystagmus were: nystagmus associated with retinal / optic nerve disease in 23 (32.4%), idiopathic or congenital motor nystagmus in 22 (31.0%), manifest latent nystagmus or latent nystagmus in 17 (24.0%), and 2 (2.8%) each associated with Chiari malformation, medication use, CNS tumor, and no diagnosis. Developmental delay was identified in 31 (43.6%) individuals, strabismus in 25 (40.3%), and amblyopia in 10 (25%). In at least one eye, eighty percent of patients presented with vision 20/40 (or equivalent) or superior.[4]

Table 1. The litelature include in this study

Author	Origin	Method	Sample Size	Result
Nash, 2017[4]	United State of America (USA)	Retrospective, population-based study	No data	This study includes population-based statistics on the occurrence of childhood nystagmus in North America as well as clinical characteristics of those affected by the condition. The majority of patients presented with normal vision and had the most prevalent presentation types, which were idiopathic and nystagmus associated with retinal or optic nerve illness. This cohort had a high incidence of developmental delay, as well as strabismus and amblyopia.
Toledano, 2015[10]	Israel	Prospective cohort study	Twenty-two patients	Both of the two individuals in this study who had CNS tumors were diagnosed with pilocytic astrocytomas of the posterior fossa at the age of 13, and both of them had unilateral nystagmus as a result of their condition. Despite the fact that tumors of the central nervous system can have a variety of clinical manifestations, nystagmus can be the first warning that anything is wrong.

Toledano, et al (2015)[10] showed there were 9 males and 13 girls with a mean age of 3.5 4.4 years; 15 were 2 years old. The tumor ranged in size from 10 6 mm to 62 29 mm. The mean duration of observation was 8.3 5.4 years. At the time of diagnosis, nystagmus was detected in 10 children (45%), all younger than 2 years of age (66.6% of the younger group); no child older than 2 years presented with nystagmus. Once present, nystagmus persisted throughout the follow-up period. During the follow-up period, there were no new cases of nystagmus in children who did not have it at diagnosis.

DISCUSSION

Nystagmus comes from the Greek words nustagmos (nodding, drowsiness) and nystazein (to become drowsy or doze). It is an involuntary, rhythmic, rapid oscillation of the eyelids. It could be slow, rapid, or a combination of the two. It can be continuous or paroxysmal, triggered by position, gaze, or head positioning. It can be distinguished from saccades, oscillations, and other fast-acting and mimicking aberrant involuntary eye movements. These movements can impair one's vision, depth perception, equilibrium, and coordination. Frequently, nystagmus is temporary, which may indicate an underlying pathology. Nystagmus may also be classified as manifest, latent, or both.[1,11]

Nystagmus is characterized by abnormal, rhythmic movement of one or both eyes and can manifest at any age. Infantile nystagmus is largely agreed upon by investigators to be nystagmus that is diagnosed within the first six months of life, which is commonly subdivided into sensory motor nystagmus (which will hereafter be referred to as nystagmus associated

with retinal/optic nerve disease), manifest latent nystagmus or latent nystagmus (MLN/LN), spasmus nutans, or idiopathic or congenital motor nystagmus (as also termed idiopathic nystagmus).[1,11]

Genetic analysis can further define cases that appear to be idiopathic. The inheritance of INS may be dominant, recessive, or X-linked. Five distinct chromosomal abnormalities (NYS1–5) are associated with INS. Mutations at locus q26 on the X chromosome have been linked to both typical clinical INS and periodic alternating nystagmus. This defect is associated with the FRMD7 gene, which is present in regions of the brain responsible for eye movement control and may play a role in neurite development. The defect is relatively prevalent among females. FRMD7 mutations have been identified in less than 10% of sporadic cases.[12]

Infantile nystagmus affected about half of the kids in this group who were identified over a 30-year period. It happened to 1 in 821 live births. Nearly 90% of the 71 people in the study had nystagmus caused by retinal/optic nerve disease, idiopathic nystagmus, or visible latent nystagmus or latent nystagmus (MLN/LN). Only 2 of the 71 people had nystagmus caused by a cancer of the central nervous system (CNS). About 80% of the patients who could see at the time of appearance had 20/40 vision or better, and about 20% had torticollis. This study group had a lot of people with developmental delays, strabismus, and amblyopia.[4]

Children have a lower incidence of acquired nystagmus (17% of all nystagmus patients) compared to adults (40% of all nystagmus patients). Diseases of the visual pathway that can cause nystagmus include craniopharyngiomas, chiasmal and optic nerve gliomas (such as those found in patients with Neurofibromatosis Type 1), and optic nerve compression caused by other types of tumors or bone abnormalities. The presence of concomitant neurological symptoms or the manifestation of vertigo, nausea, and headaches as a result of high intracranial pressure can be found in a significant number of individuals who have neurological nystagmus.[13]

Although there are several ideas that seek to explain the development of infantile nystagmus, the underlying mechanism that causes infantile nystagmus is not entirely understood. Fixation, saccades, the optokinetic reflex, the neural integrator, or the pursuit eye movement systems are the ones that are most likely to be affected by the condition. The genesis of the disorder in all of its complexity in an oculomotor movement system that mostly retains intact saccades, pursuits, and VOR movements outside of the nystagmus has not been explained by a single explanation as of yet.[12,14]

Both of the two individuals in this study who had CNS tumors were diagnosed with pilocytic astrocytomas of the posterior fossa at the age of 13, and both of them had unilateral nystagmus as a result of their condition. Despite the fact that tumors of the central nervous system can have a variety of clinical manifestations, nystagmus can be the first warning that anything is wrong.[10] When evaluating a kid or infant who has been diagnosed with nystagmus, it is essential to ascertain the age at when the condition first appeared, in addition to the child's birth, developmental, medical, and family history.[15]

When diagnosing the cause of INS in children, more recent high-resolution imaging modalities like optical coherence tomography (OCT) are being used more and more. The ability to use imaging to complement the clinical evaluation is significant because individuals are frequently misdiagnosed as having idiopathic INS when, in reality, there is an underlying ophthalmic disease, and foveal maldevelopment can be discovered on OCT. The ability to use imaging to augment the clinical evaluation is important because of this. When the start of nystagmus is after three months, is not associated with an underlying sensory deficiency, or is associated with optic nerve hypoplasia, neuroimaging should be examined.[16,17]

Vision-specific health-related quality of life in children with vision impairment is poorly understood. The few studies that do exist have evaluated groups of children with diverse causes of vision impairment, of which only some are associated with nystagmus. The majority of research on the effects of nystagmus on quality of life and visual function has been conducted on adults. Clearly, more research is required in this field. Using parallel focus groups of children with vision impairment and their caregivers, DeCarlo et al. examined the effects of pediatric vision impairment on quality of life. Nystagmus was present in 76% of the children who participated.[18]

CONCLUSION

Nystagmus is involuntary, conjugate, rhythmic eye movement. Infantile nystagmus syndrome (INS) causes most child nystagmus. Understanding the treatment choices for this lifelong illness is crucial. This review discusses nystagmus genesis, psychosocial and functional implications, and current management methods, including optical, pharmaceutical, surgical, and rehabilitative approaches.

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