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Nystagmus

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Abstract

Nystagmus is an involuntary rhythmical movement of the eyes. The cause of nystagmus is a disruption in the afferent, central or efferent parts of the eye movement system. If it happens in the first few months of life during the sensitive period of visual development, it is most often a case of infantile nystagmus. On the other hand, the majority of nystagmus in adult age is caused by some neurological disorder, and it is usually called acquired nystagmus. The important role of an ophthalmologist is to recognize the origin of nystagmus. Acquired forms are usually caused by some neurological disorders and do not belong in our field of treatment. However, most of the nystagmus types in a child's age require ophthalmological treatment. When we have a child with nystagmus, we have to enable the development of the visual system and help fixation and fovealization by the dampening of nystagmus. If the reason of nystagmus is of ocular origin, we have to treat the underlying disease. Optical treatment by glasses, contact lenses or magnifying devices is usually reasonable. In some cases when the patient has abnormal head posture, it is possible to treat nystagmus by surgery. Some medications are used in several types of nystagmus as well as some new developing treatments.

Keywords: nystagmus, infantile nystagmus, acquired nystagmus, spasmus nutans, albinismus, artificial divergence, Anderson surgery, Kestenbaum surgery

1. Introduction

When examining the patients with nystagmus, we should start with the medical history. One of the most important questions, especially in a child's age, is at what age nystagmus started (infantile, benign form starts no later than 3 months of age). Nystagmus usually consists of slow pursuit movement or drift which is followed by a fast (jerk) or slow (pendular) movement of refixation. Though the first slow movement is pathological, nystagmus is usually named after the second, fast refixation movement. When the second movement is fast, nystagmus is called "jerk", and in the cases when second movement is slow, nystagmus is called "pendular". When describing nystagmus we usually describe the direction of the movement that can be horizontal, vertical, rotatory or chaotic as well as the magnitude of amplitude and frequency. It is important to emphasize that in cases when the movements of both eyes are asymmetric, meaning that eyes do not have the same amplitude, frequency or direction (dissociated nystagmus) or eyes moving in opposite directions (disconjugate nystagmus), we have to consider that as an alarming sign of central cause of nystagmus and send the patient immediately to a neurologist. Besides doing the clinical analysis of nystagmus, we can record eye

movements by electrooculography. By using eye movement recording tests, we can analyze eye movements in detail and help better analysis of nystagmus in clinical research.

As ophthalmologists we have to pay special attention to eye conditions that are accompanied or caused by nystagmus. During a complete ophthalmological clinical examination of the eyes in the children with nystagmus, it is important to pay attention to the preferred head posture and associated eye abnormalities and take care that monocular visual acuity can be different if one eye is closed, so it is wise when performing visual acuity tests to dampen instead of close the non-tested eye [1, 2].

2. Types of nystagmus

Nystagmus is a complicated disease that most of us do not see every day in our offices.

According to its origin, nystagmus is usually divided in physiological, infantile and acquired [3]. An ophthalmologist has an important role to recognize the origin of nystagmus. Acquired nystagmus is usually caused by some neurological causes, and this complex disorder is in the field of neurologists as well as ear, nose and throat (ENT) specialists and not in our field of treatment. However, we should recognize the type of nystagmus and help localize the disruption. As ophthalmologists, we usually treat nystagmus in childhood. Most of the nystagmus types in a child's age are benign, and they do not require additional neurological workup [4, 5].

After medical history and clinical examination in the child with nystagmus, we can perform measurements of nystagmus by electronystagmography, as well as measurements of the afferent part of the system with visually evoked potential (VEP), optical coherence tomography (OCT) or electroretinogram tests.

2.1 Physiological nystagmus

Since the visual acuity is degraded by image slip across the retina more than 2–3°/s, some nystagmus-like movements of the eyes are physiological and unconsciously used in everyday activity (Table 1).

Physiological fixational nystagmus	Nystagmus-like movements
Optokinetic	Microsaccade
Gaze evoked	Microtremor
Caloric	Slow drift
Rotatory	

Table 1.
Physiological fixational nystagmus and nystagmus-like movements.

There are several systems that are connecting the eyes (afferent part), structures in the brain and eye movement system (efferent part). Each of them is responsible and important in the cases when our body is moving or rotating or when we are fixing a moving target (Table 2) [3].

	Afferent visual system	Vestibular system		Neural integrator
Function	Provides image to guide eye movements	Moves eyes in response to self-motion		Maintain eccentric gaze in the presence of moving target
Anatomy	Retina Optic nerve Optic chiasm	Central	Peripheral	Medial vestibular nucleus Nucleus prepositus hypoglossus Flocculus Interstitial nucleus of Cajal

Table 2.
Eye movement systems.

2.2 Nystagmus in childhood

Benign nystagmus in childhood can be divided in several forms:

Idiopathic infantile nystagmus

Ocular/sensory nystagmus

Latent (manifest/latent) nystagmus

Spasmus nutans

Typical features of each form are listed in **Table 3**.

Type of nystagmus	Infantile nystagmus syndrome	Sensoric nystagmus	Manifest/latent nystagmus	Spasmus nutans
Onset of nystagmus	3–4 months	3–4 months	3–4 months	6 months–2 years
Additional eye findings	Early esotropia syndrome	Congenital cataract Cloudy cornea Albinism Congenital macular disease Optic nerve hypoplasia	Early esotropia syndrome	Head nodding
Visual acuity	Variable	Poor	Variable	Good
Prognosis	Recedes by the age of 8–9 years	No change	No change	Recedes in 1–2 years
Treatment	Visual aids, prisms, surgery	Treatment of underlying disease, visual aids	Visual aids, surgery	None

Table 3.
Benign nystagmus of childhood.

2.2.1 Idiopathic infantile nystagmus

Idiopathic infantile nystagmus is a primary motor dysfunction of unknown origin with no ocular pathology present. In some cases, it can be connected to some genotype but without a clear phenotypic pattern [6, 7]. Inherited forms are usually X-linked form in the FERM domain on Xq26.2 chromosome [8] or autosomal dominant situated at the chromosome 6p12 (NYS2) [9], 7p11 (BYS3) or 13q (NYS4) [10].

Idiopathic infantile nystagmus is probably present from birth but typically becomes noticeable in the first several months of life when a child starts to fixate. If it develops later, we have to suspect other kinds of nystagmus. It starts as a pendular form, later transforms into the jerk type and usually recedes spontaneously up to the 9th year of life, although it is usually present for lifetime. Idiopathic infantile nystagmus is usually horizontal and symmetrical but can have a rotatory component as well. Although it is typical that patients with infantile nystagmus do not have oscillopsia, they sometimes have head shaking or nodding, and it is also typical that nystagmus disappears during sleep. Some recent studies of patients with infantile nystagmus showed downregulation of the visual cortex which is responsible for motion processing (MT/V5 area) [11]. Typically patient with this kind of nystagmus does not have optokinetic nystagmus, and vestibulo-ocular reflex is disrupted [12]. Infantile nystagmus often (even up to 50%) comes with esotropia and is combined with some vertical movement disorders (congenital form of fourth muscle palsy or dissociated vertical deviation). Such a combination is then called infantile esotropia syndrome.

Infantile nystagmus is typically increasing when attempting to fixate a small visual object. The majority of children can dampen eye movements in convergence or some other eccentric direction of gaze. In the preferred position, sometimes called “null or neutral zone”, nystagmus is dampening, so the fixation and fovealization are better, and children often have compensatory head position [13, 14]. In patients with infantile nystagmus, the development of the visual acuity is variable. Visual acuity can be 20/20 but sometimes it can be severely reduced.

2.2.2 Ocular/sensory nystagmus

This is a kind of nystagmus which develops in cases when, during the early visual development (the first few months of life), the development of vision is not possible because of anatomical changes or organic defects in the eye. Disorders of the eye can be obvious, but sometimes they are subtle and not easy to define [15]. The cause of sensory nystagmus is inadequate image formation in fovea which results in disruption between afferent and efferent system and causes disturbances of oculomotor control. The most common causes are diseases of the eye such as congenital cataract, albinism, corneal opacities as well as developmental abnormalities of the optic disc and retina such as Leber’s amaurosis, achromatopsia or stationary night blindness [16]. Specific form of sensory strabismus is unocular nystagmus in deep amblyopia (Heimann-Bielschowsky phenomenon).

2.2.2.1 Congenital cataract

Congenital cataract is opacification of the lens present at birth and may be responsible for amblyopia, sensory nystagmus and strabismus. The incidence varies between 2 and 4 in 10,000 worldwide [17].

The vast majority of congenital cataracts are bilateral, roughly between 60 and 70%. Identifiable cause of the cataract can be found in only half of them. Unilateral cataracts are mainly sporadic, found in otherwise healthy infants [17]. When they are associated with other local or system abnormalities, unilateral cataracts are usually found in eyes with other ocular abnormalities, and bilateral cataracts are mainly associated with system disorders (**Table 4**).

Not every lens opacification is equally important for normal vision development. The most sight threatening is if opacifications are central, more than 3 mm, located from central to the posterior parts of the lens. While sensory nystagmus usually develops in bilateral congenital cataracts, strabismus may occur in unilateral and bilateral cataracts.

Cataract	Unilateral	Bilateral		
Associated abnormality	Persistent foetal vasculature	Genetic	Genetic	AD
	Posterior lenticonus/lentiglobus	malformations	mutations	AR
	Anterior segment abnormalities			X-linked
	Chorioretinal coloboma		Chromosomal abnormalities	21 trisomy 18 trisomy 5p partial deletion
		Metabolic disorders		Galactosemia Wilson's disease Hypocalcemia DM
		Intrauterine infections		Rubella*, Rubeola HSV, CMV, HZV, EBV Toxoplasmosis Syphilis

**Rubella is the most common intrauterine infection causing congenital cataract; DM, diabetes mellitus; HSV, herpes simplex virus; CMV, Cytomegalovirus; HZV, herpes zoster virus; EBV, Epstein-Barr virus.*

Table 4.
 Associated abnormalities in unilateral and bilateral cataracts.

Nystagmus is found in at least 24% of eyes with bilateral cataract, being thought to be a poor predicting factor for developing normal visual acuity [18].

As the risk of amblyopia is the greatest in the earliest months of life, it is of crucial importance to detect the cataract as soon as possible. That is why all infants must be screened for congenital cataracts immediately after birth and from 6 to 8 weeks of life. Screening is based on examination of red reflex. Any shadow in the reflex, absence or whitening of the reflex is an indication for complete ophthalmology examination [19]. On ophthalmology exam, it is essential to have a clear picture of fundus which is not blurred by opacification of the lens [17]. The more obscured fundus means that the vision development is more affected, and that determines timing of the surgery.

In cooperation with a pediatrician, set of laboratory examinations should be performed, even in cases without the presence of notable system abnormalities. If dysmorphic features are present, a genetic test should be suggested. Laboratory tests that should be done are TORCH titers; venereal disease research laboratory test (VDRL test); serum levels of calcium, phosphorus and blood glucose; and urine analysis for reducing substances (raised in galactosemia), galactokinase (raised in Fabry's disease), amino acids (raised in Lowe syndrome), calcium and phosphorus [19].

2.2.2.2 Albinism

Albinism is a disorder which is characterized by reduced pigmentation of the skin, hair and eyes caused by inborn defects in melanin biogenesis and distribution. The defect can be present as an isolated form or, less frequently, as a part of the syndromes (Chediak-Higashi, Hermansky-Pudlak, Waardenburg) [20]. Although four genes are known to cause autosomal recessive form, it seems that, only in one-third of the patients with albinism, mutations in known genes are confirmed [21]. If the lack of pigmentation is present in the skin, hair and eyes, the disorder is called oculocutaneous albinism (OCA); if the hypopigmentation is present only in the eyes, it is called ocular albinism (OA) [22]. Pigments have multiple functions in the development and protection of the visual system. Although the exact molecular mechanisms are not yet completely understood, it is well known that pigment is crucial in some critical steps of the visual development as well as a factor of

protection of damaging light and cellular protection by being a trap for free radicals [23]. Important features of ocular albinism that causes reduced visual acuity are macular hypoplasia and abnormal decussation of the visual pathways [24, 25]. Because of the reduced visual acuity in patients with albinism amblyopia, nystagmus and strabismus often develop. Macular hypoplasia is the most important factor responsible for the reduced visual acuity in patients with albinism and is easily diagnosed by inspection (lack of foveal pit, abnormal growth of blood vessels) and confirmed by OCT (optical coherence tomography). Another important feature is the abnormal decussation of the visual pathways. In a normal eye, one half of the nerve fibers decussate to the contralateral side. On the other side, in patients with albinism, 75–85% of fibers project to the contralateral side [26]. The reason for that is lack of melanin in the specific time of development when melanin and its precursors have an important role in linking reception in the retina and perception in the brain [27].

With the clinical picture of the nystagmus alone, it is not possible to tell the difference between this kind of nystagmus and idiopathic infantile type. However, in this type visual acuity is severely reduced with no chances of improvement in contrast to the idiopathic form where the visual acuity can be normal. This type of nystagmus does not reduce during the years.

2.2.3 Latent nystagmus

Latent/manifest nystagmus is benign, jerk kind of nystagmus that starts early in the childhood. Although this type of nystagmus is bilateral and conjugated, the main characteristic is that the nystagmus is not visible (or much less visible) when both eyes are open. It increases when one eye is closed. Other typical feature of latent nystagmus is that it changes direction. The fast phases are always towards the open eye. When the right eye is closed, it beats left, and when the left eye is closed, fast phase is right [28]. Sometimes this kind of nystagmus is called fusion defect nystagmus (FDN) because it is present (or more pronounced) with one eye closed when fusion is disrupted [29]. This kind of nystagmus is usually combined with other anatomical eye abnormalities, and most often it is part of the early esotropia syndrome [30].

2.2.4 Spasmus nutans

Spasmus nutans is a benign nystagmus of childhood which is dissociated (different amplitudes, directions or frequencies between two eyes). The frequency is usually high, movement pendular, with a small amplitude, and disconjugated oscillations. The movements can be horizontal, vertical or torsional. It typically starts later than the idiopathic form, usually at the age of 4–12 months. If it starts after 3 years of age, the possibility of an intracranial tumor is strong. It is often preceded by the head nodding several months before the appearance of the nystagmus itself. Spasmus nutans is a specific form that can be a problem for diagnostics since there is no certain clinical sign that can differentiate this kind of nystagmus from the nystagmus caused by neurological problems (tumors in diencephalon) [31]. It is necessary for this kind of nystagmus to perform a complete neurological and endocrinological workup. Spasmus nutans typically disappears spontaneously in the 4th year of life [31].

2.3 Acquired nystagmus

As mentioned in (Table 2), several systems working in synchrony are responsible for involuntary movements of our eyes. The disorder that creates

Affected system	Afferent visual system	Vestibular system	Neural integrator
Etiology	Neuroblastoma Optic nerve glioma	Tumors (brainstem) Stroke (brainstem) Degenerative diseases Drugs	Cerebellar diseases Brainstem diseases Drugs

Table 5.
Acquired nystagmus.

pathological nystagmus can be situated in some parts of these systems or in the surrounding parts of the brain, brain stem and cerebellum. Most often the reasons are strokes or mass lesions, trauma, multiple sclerosis, some malformations and drugs (Table 5).

Another important characteristic that usually makes difference between acquired and benign forms is that acquired nystagmus is usually combined with other neurological signs like nausea, vomiting, headache, vertigo or tinnitus. In some cases the type or direction of the nystagmus can help us localize the place of lesion. For example, vestibular system is responsible for moving eyes in the opposite direction of the moving of the head. Both sides of system work in balance. When one side of the system is damaged, balance is lost, and the eyes will beat towards the not affected side and do not change side when the gaze changes direction. It is typical for this kind of disruption that nystagmus is increasing when the patient is not fixing. Lesions of the peripheral part of vestibular system (labyrinth) are accompanied by ataxia, vertigo and other signs of disturbances of the vegetative system. On the other hand, in the cases when damage is in the brainstem, often involving vestibular nuclei (fasciculus longitudinalis medialis), the direction of nystagmus is changing with the direction of the gaze, with the amplitude increasing when looking at the affected side. The intensity of nystagmus is increasing with fixation. In the cases of cerebellum diseases, nystagmus is increasing with fixation, more pronounced with the bigger amplitude and slower frequency when the gaze is towards the affected side. Vertical nystagmus is usually of central origin, and in some cases, it can be caused by excessive sedative medications intake. Acquired nystagmus can be a life-threatening condition which sometimes requires urgent neurological treatment [3]. Although in child's age the incidence of acquired nystagmus is smaller than in adult groups (17% in children compared to 40% in adult groups) [4, 5]. In our clinical work, it is of crucial importance when examining a child with nystagmus to notice the signs that are warning us that nystagmus is neurological in its origin, which means that the cause which is central does not require our treatment but needs neurological examinations, imaging and treatment [1].

Signs for neurological cause of nystagmus are:

- Neurological symptoms (vertigo, nausea, headache, vomiting)
- Existence of the additional signs on the eyes (oedema of optic nerve, RAPD)
- Nystagmus which is disconjugated or dissociated
- Oscillopsia
- Existence of optokinetic reflex
- Appearance of nystagmus after the 4th months of age [1]

3. Treatment

In the cases when nystagmus is acquired, it is necessary to treat the reason that caused nystagmus. That treatment is usually in the hands of neurologists or neurosurgeons. When we have a child with some type of benign nystagmus, the primary goal is to improve proper development of the visual system which is in the hands of ophthalmologists. When the baby is born, the visual system is not fully developed, and it needs proper stimulation at the proper time for the cells in visual parts of the brain to develop to their full potential. We can say that a child has to learn how to see. Children with nystagmus often have refractive errors such as astigmatisms, myopia or hyperopia, given prescription can enable better development of the visual system and with improvement of fixation and fovealization nystagmus can dampen. In the cases of very poor visual acuity, some magnifying visual aids can be helpful [32].

Often children with nystagmus can have abnormal head position to enable better fixation so the optical axis of the glasses is not the same as the visual axis of the eye. In cases like that it is wise to prescribe contact lenses. The reason of the preferred position of the head is so called null zone of nystagmus. Certain types of nystagmus have eye position in which nystagmus is less pronounced. By abnormal head position, the child is positioning the eyes in the preferred position where nystagmus is dampening. Prescribing the prismatic correction (base towards the head turn) can shift the null zone and correct head position. Prism correction can correct only mild head turn and the does not have permanent effect so cannot be prescribed like a definite therapy. Bigger amounts of prism correction are heavy and cause chromatic aberration, and Fresnel prisms will degrade visual acuity.

In the cases when the preferred position is bigger than 10° , there is a possibility to perform surgery on the eye muscles to move the eyes in the direction of the head turn, in order to shift the dampening zone from the decentralized position to straight ahead, to enable the patient a better and easier fixation. When considering a possibility of surgery, we have to think that some kinds of nystagmus spontaneously dampen by the age of 8–9 years, so the surgery can be planned in older children.

Most commonly performed surgeries are:

Anderson type of surgery

Kestenbaum type of surgery

Artificial convergence

Y-splitting

Posterior suture (Faden surgery)

Anderson type of surgery consists of recession of horizontal muscles of both eyes. Which muscles will be operated depends on the position of the eye and the head. Anderson started with the recession of one muscle on each eye with the idea that with the recession of the muscle he will weaken the tension and contact between the eye and the muscle which can help in the dampening of the nystagmus [33]. But the main effect of this kind of surgery is shifting the gaze direction in to the dampening zone. His original idea was recession of yoke muscles of 4 mm [33]. After his idea many modifications are made from other authors with different amounts of recession but taking into consideration that the amount must be symmetrical on both eyes in order to prevent induction of strabismus. The advantage of this method

is that we are not only shifting eyes in the right direction but weakening the contact between the eye and the muscle and by that helping to dampen nystagmus.

In some cases, it is impossible to correct the head positioning only with recessions, so we have to add resections. That kind of surgery is named after Kestenbaum who published his method at the same year as Anderson [34]. Depending on the position of the head, a combination of recessions and resections is performed. This kind of surgery can be done on vertical and oblique muscles in cases when the patient is lifting, lowering or tilting the head [35]. During the years, many modifications of these surgeries have been done, mostly with recommendations for bigger amounts of surgery. Originally, Kestenbaum proposed recessions and resections of 5 mm; one of the most popular recommendations is done by Parks “5,6,7,8 procedure” with 13 mm of surgery in each eye [36]. Calhoun, Harley, Nelson and Pratt-Johnson suggested “augmented Parks surgery”; some of them prefer up to 10 mm of recess resect surgery [37, 38].

A different type of surgery is recommended for patients who are turning the head when looking at far distance and having their head straight when reading. The reason for dampening of nystagmus is convergence that they are using for near work. In such patients, we can perform a procedure called “artificial divergence”, meaning that we create a latent divergence by surgery so the patient has to use convergence when looking at far distance (like during reading at near) and by that is dampening the nystagmus. When performing this type of surgery, we have to be sure that the patient has binocularity (possibility to use both eyes together); otherwise we will not achieve the goal of the surgery. It is always wise to simulate the wanted postoperative position by using prisms before the surgery. In the case of artificial divergence, we can check the amount of wanted divergence by putting prisms on both eyes and checking whether the patient is converging or accommodating and by that better estimate the amount of the needed surgery [39].

Some patients are changing their preferred head position, sometimes turning left and other time right with dampening nystagmus when the eyes are in convergent position. Often they do not have binocularity as the group mentioned previously, so we cannot use their active convergence to dampen nystagmus. In cases like this, Y-splitting of the medial rectus can be performed. By splitting both medial recti in two half up to 15 mm from the insertions, we create two arms of one muscle and fixate them away from each other. The exact position of placing the respective arms is calculated using a mathematical model in which axial length and angle of additional squinting must be added to the formula. By this kind of surgery, we block action of medial muscle only in adduction and by that we enable the patient to have convergence without active power of binocularity [40].

Similar effect has posterior fixation surgery or Faden surgery. When performing this kind of surgery, we suture the muscle to the globe at 15 mm behind the limbus and with that block the action of the muscle in the desired position [41]. Both kinds of surgeries are usually used on medial rectus muscles but can be used on other rectus muscles of the eye if we want to reduce the action of the operated muscle [42].

Nowadays we have some medications that are used for treating nystagmus (memantine, gabapentin, baclofen 4, aminopyridine, etc.), but because of a number of side effects, they are usually not used for children [43]. Some modern therapies like biofeedback (making the patients aware of the eye bobbling by sound or touch, and by that teach the patient to control nystagmus) are sometimes performed, but since there is no permanent effect, it is not widely used [44, 45].

Recently some proprioceptive nerve endings have been found at the place of the insertion of the eye muscle at the globe. Some authors advocate the idea that the cutting of the eye muscle at their insertions with reattaching or giving some

medications (brinzolamide) that act on these endings can change signals and re-boost ocular motor connection and with that dampen the nystagmus [46, 47].

Conflict of interest

The authors declare no conflict of interest.

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