1-1-1986

Classification of nystagmus and field observation at two blind schools

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Abstract
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Degree Type
Thesis

CLASSIFICATION OF NYSTAGMUS AND FIELD OBSERVATION AT TWO BLIND SCHOOLS

A SENIOR THESIS
Presented to
The Faculty of the College of Optometry
Pacific University

In Partial Fulfullment
of the Requirements for the Degree
Doctor of Optometry

by
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January 1986

ACKNOWLEDGEMENTS

The authors wish to recognize and thank Dr. Alfred Furie, for his role as an advisor to the project. The authors would also like to express their gratitude to the respective staffs at each of the participating blind schools.

CLASSIFICATION OF NYSTAGMUS

Nystagmus is defined as a rapid, repetitive, involuntary, oscillatory movement of the eyes. The condition can be classified by its ,etiology, pattern, and whether it is pathological or physiological. This paper will deal with pathological nystagmus only.

The two patterns of nystagmus that are observed are pendular and jerk. Pendular is an undulatory movement of equal speed and amplitude in each direction. Jerk consists of a biphasic rhythm with a slow movement in one direction followed by a rapid saccadic return to the original position.[1] These two patterns may occur by themselves or mixed. When the pattern is mixed one can observe pendular nystagmus in one position of gaze and jerk nystagmus in another.

CONGENITAL

Any nystagmus observed at birth or in early infancy is classified as congenital.

Sensory defect nystagmus which can result in pendular nystagmus, is a result of impairment to any part of the visual pathway. Head oscillations in opposite directions of the nystagmus often occur. The causes are high refractive errors, myopia, congenital or traumatic cataracts, corneal dystrophies, congenital glaucoma, albinism, aniridia, achromatopsia, optic atrophy, and lesions of the posterior pole.[2]

Motor defect nystagmus which usually results in jerk nystagmus, has a fast component in the direction of gaze. It is probably due to a lesion of the brainstem. A position of relative rest is always present.

Latent nystagmus results in a jerk pattern, and is only manifest when one eye is covered. According to Von Noorden and Daroff, Troost, and Dell'Osso, the fast phase is towards the uncovered eye.[2,3] According to Duke-Elder and Vaughan and Asbury, the fast phase is towards the covered eye.[1,4] The cause is unknown.

SPASMUS NUTANS

This is a rare condition occurring in infants between the ages of four and eighteen months, and usually lasts three months to two years. One observes ocular oscillations, head nodding, and torticollis. The nystagmus is generally binocular and pendular. The direction can be either horizontal, vertical, or rotary. The cause is unknown.

VESTIBULAR NYSTAGMUS

This jerk nystagmus is associated with dysfunction of the vestibular system. The slow phase is in response to an impulse which originates in the semicircular canals, and the fast phase is a corrective movement.[4] The movement may be horizontal, rotary, or vertical. This can be caused by lesions to the brainstem or end organ in conditions such as encephalitis, multiple sclerosis, syringobulbia, poliomyelitis, and thrombosis of the

posteroinferior cerebellar artery. Other causes include Menier's disease, destruction of a labyrinth, vertigo, tinnitus, and deafness.

GAZE NYSTAGMUS

Of all the types of nystagmus, this is most often seen clinically. It occurs when the eye is in an eccentric position with the fast component in the direction of gaze. The slow component is a drift of the eye back to the primary position. When the condition is coupled with deficient smooth pursuits, it is indicative of cerebellar dysfunction. The condition can also be brought on by sedative or anticonvulsant medication. Myasthenia gravis is another cause.

VOLUNTARY AND HYSTERICAL NYSTAGMUS

Both of these are similar to one another. The pattern seen is a rapid horizontal nystagmus of high frequency and low amplitude. This can only be sustained for a few seconds. Voluntary nystagmus is employed as a trick, and hysterical nystagmus is involuntary and associated with anxiety neuroses.

AMAUROTIC NYSTAGMUS

Those who have been blind for a long time will usually have this. It is mostly pendular and rarely jerky.

AMBLYOPIC NYSTAGMUS

It may be due to a compensation for a central scotoma or an ataxia of the extraocular muscles from the absence of foveal stimulation. This is usually developmental in origin and is due to a defect in central vision. The causes are amblyopia in albinism or optical albinism, achromatopsia and congenital or infantile ocular anomalies in the media or retina.[1] This condition is comparable to congenital sensory defect nystagmus discussed earlier.

DISSOCIATED NYSTAGMUS

This is characterized by an asymmetry in either amplitude or direction of the two eyes. The causes include lesions to the medial longitudinal fasciculus, lesions associated with multiple sclerosis, and posterior fossa lesions.

ROTARY NYSTAGMUS

This is also known as torsional nystagmus. In vestibular end-organ dysfunctions there is a rotary component combined with vertical and horizontal nystagmus. If the amplitude is small this is associated with a medullary lesion. A larger amplitude reflects a congenital nystagmus. If the rotary nystagmus is acquired, then it is due to thalamic involvement.[3] Refer to vestibular nystagmus.

DOWNBEAT NYSTAGMUS

In the primary position the fast phase is in a downward direction. This is brought on by brainstem disease or drug intoxication. Specific examples of brainstem disease include Arnold-Chiari malformations and parenchymal cerebellar disease.

UPBEAT NYSTAGMUS

In the primary position the fast phase is in a upward direction. There are three types of upbeat nystagmus classified according to amplitude. The first type is of large amplitude and caused by a lesion in the vermis of the anterior cerebellum. The intermediate type is a manifestation of Wenicke's encephalopathy prior to the institution of thiamine therapy.[3] The last type is of small amplitude and suggests intrinsic medullary disease.

OCULAR BOBBING

One will see fast downward jerks in both eyes which is followed by a slow drift back to the mid-position. This is usually observed in comatose patients that have destruction of the pons.[3]

SEE-SAW NYSTAGMUS

This has an alternating oscillation in which one eye goes up while the other eye goes down. The causes include parasellar tumors, head trauma, and midbrain infarction which result in a bitemporal hemianopia.

TREATMENT

SURGICAL

In the mid 1950's, Alfred Kestenbaum of New York and J. Ringland Anderson of Melbourne developed nystagmus surgery.[1,5]

Anderson's procedure involves resectioning the two lateral recti in order to rotate the eyes toward the side of the greatest nystagmus.[5]

Kestenbaum's procedure is similar to Anderson's but includes resectioning of all four horizontal recti.[5]

The objective of the surgery is to shift the neutral zone to the primary position of gaze. The neutral zone is the position in which there is the least nystagmus.

If no heterotropia is present, care must be taken not to create a strabismus as a result of the surgery. The amount of resectioning depends on each individual case, but certain guidelines have been given. The maximal medial rectus resectioning is 5 mm. To balance the lateral and medial recti, the lateral rectus is resectioned 1 mm more than the medial rectus, according to Anderson.[6]

Kestenbaum believes that both the medial and lateral recti should be resectioned 5 mm each.[6]

The amount of resectioning of an individual muscle depends on the magnitude and direction of the head turn. For example,

Dale recommends that if the face turn is to the right, the right medial rectus should be resectioned 5-6 mm, the right lateral rectus 8-10 mm, the left lateral rectus 7-8 mm, and the left medial rectus 6-7 mm.[7]

If there is a head tip rather than a head turn, the inferior and superior recti are each resectioned 4 mm.[7]

Complications of the surgery include diplopia, induced strabismus, and a deviation in the opposite direction in someone who already has strabismus. According to Dell'Osso and Flynn, these complications are quite common and often require an additional operation within three months of the original operation.[8,9]

Since these procedures for nystagmus correction have such variable results, considerable research and refinement of the techniques, needs to be done.

NONSURGICAL

PRISMS

Prisms will shift the neutral zone to the primary position of gaze to eliminate head tilt. Dell'Osso states that the patient will require less effort to fixate and at the same time will increase their visual acuity.[10] Prisms may also be used to find the neutral zone in order to calculate the amount of resectioning required in surgery.[11]

The base orientation of the prisms depends on the fusional power of the patient. If the fusional power is good, base out

prisms are used. If the fusional power is poor, prisms are placed with bases in the same direction with an equal power. The bases are oriented in the opposite direction to the preferred position of gaze.[12]

LOW VISION AIDS

Nystagmus due to low vision is often corrected with low vision aids. Before prescribing an aid, it must be decided if the nystagmus is due to the low vision or if it is occurring independently.

Three interactions between magnification, visual acuity, and nystagmus are possible. The first is that magnification may improve visual acuity in the presence of nystagmus. The second is that magnification may have no effect on visual acuity in the presence of nystagmus. The third is that magnification may reduce visual acuity in the presence of nystagmus.[13]

If the nystagmus is primary and magnifies the image, it will only magnify the oscillation and decrease the visual acuity. When the nystagmus and low vision are independent, a limited improvement in visual acuity will be observed. If the nystagmus is a result of the patient's impaired vision, magnification will decrease the oscillation and increase visual acuity.

Before prescribing a low vision aid, different magnifications should be tried to see which will work the best for the patient.

CONTACT LENSES

As discussed before, nystagmus is often caused by aniridia or albinism. A person with these conditions can benefit from wearing a contact lens, because the incident light which enters the eye can be more easily controlled.[14]

Corneal irregularities can also cause a sensory defect type of nystagmus. Hard contact lenses tend to eliminate much of the irregularity of corneal scarring. As a result, visual acuity is increased, and the nystagmus is lessened.

For those patients which have a high refractive error, spectacle lenses can cause image distortion, induce prism effects, and because of this the nystagmus movement is not decreased. In these cases, contact lenses are beneficial because they do not present any of the disadvantages of spectacle lenses mentioned above.[12] Contact lenses also move with the eye which helps to increase visual acuity.[15]

According to Abadi, the amplitude and frequency of the nystagmus can be diminished if contact lenses are worn. The contact lenses also require additional vergence and accommodative effort.[14]

The most frequently used type of contact lens is a hard gas-permeable corneal contact lens because it gives the maximal visual benefit. Younger patients are initially fitted with daily-wear soft contact lenses to allow them to gain confidence in handling. They are then changed to a hard gas-permeable lens.[15]

AFTERIMAGE TREATMENT

Stohler used an electronic flash to produce an afterimage. The device used is similar to the euthyscope or projectoscope. An afterimage is placed on the patient's fovea. For the patient to discern both a positive and negative afterimage, overhead illumination is turned off and a blinking light is placed behind the patient. The patient then observes the afterimage oscillating according to the type of nystagmus present. This will make the patient aware of the nystagmus movements. The patient must learn to move the head to a position where there is no nystagmus movement perceived.

Afterimage treatment has been effective for a small number of patients. More research needs to be done in this area because of the small number of patients treated.[16]

BIOFEEDBACK

Biofeedback therapy refers to the method of learning to control one's bodily and mental functions with the aid of a visual or auditory display of these functions.

Auditory biofeedback was first developed by Abadi, Carden, and Simpson. Their goal was to improve sensory and motor function in patients with congenital nystagmus.[17] The method used consisted of varying the tone frequency in accordance with the amplitude and direction of the horizontal eye position. The patient was asked to maintain the pitch at a single level. The therapy resulted in an average decrease of 28% for the amplitude, 29% for the

frequency, and 41% for the slow phase velocity. Visual sensitivity was also increased.

A more recent method of auditory biofeedback has been developed by Ciuffreda, Goldrich, and Neary. The horizontal eye position is monitored by a photoelectric technique, with the patient hearing the output through a loudspeaker. The quality of the tone reflects the steadiness of the patient's gaze, and the tone must be kept as steady as possible. The results showed that there was decrease in the amplitude of 82%, 86% in the slow phase velocity, and 34 % for the frequency.[18]

Kirschen also uses an auditory biofeedback method. An infrared eye movement monitor detects horizontal eye position with respect to the fixation target. The patient hears a sine-wave tone in his/her right ear when the eyes are to the right of the target, and a square-wave tone is heard in the left ear when the eyes are to the left of the target. No tone is heard when the patient is viewing the target; therefore, the patient is instructed to make all tones disappear.[19]

A method of visual biofeedback developed by Goldrich involves the use of the Goldrich Contour Rotator (GCR). The patient is made aware of the nystagmoid movement by viewing the changes in the textural contour of the illuminated grid. With steady eye position, the patient will perceive a uniform contour.[18,19]

FIELD OBSERVATION AT TWO BLIND SCHOOLS

In order to view the different types of nystagmus, we visited two blind schools in April of 1985.

At the first blind school, the staff went through the records to locate the students that had nystagmus previously diagnosed. Two or three students were observed for less than a minute in informal situations. The students were between the ages of five and twenty-one. We saw approximately half of the sixty-two students enrolled at the school.

During the visit to the second blind school, we observed thirty-eight of the fifty-five enrolled students. For this visit, the records were not reviewed for cases of diagnosed nystagmus. We observed each student individually in the classrooms. The age range was the same as in the first blind school.

The two prevalent types of nystagmus seen in both schools were pendular and jerk. Almost all students exhibited either pendular or jerk rather than a combination of both. Of the eighteen cases of nystagmus in the first blind school, fifteen were pendular and three were jerk. There were fifteen cases of nystagmus in the second blind school, including twelve pendular and three jerk.

Since our contact with the students was limited, it is possible that a different type of nystagmus could be observed in other situations. It has been stated that pendular nystagmus is possibly related to a sensory deprivation, and jerk nystagmus to a

motor defect.[1,2]

We feel that the prevalence of pendular nystagmus in the students we observed could be due to their low vision status.

We felt that this field study was worth while because it enabled us to observe different types of nystagmus first hand. We would encourage further research in the area of nystagmus and its treatment.

TYPES OF NYSTAGMUS

OCCURRENCES	PENDULAR	JERK	TOTAL PATIENTS
FIRST BLIND SCHOOL	15	3	18
SECOND BLIND SCHOOL	12	3	15
TOTAL PATIENTS	27	6	33

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