Positional Nystagmus

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In the latter part of the last century Ewald (1892)[1], Mach (1875)[2], Breuer (1874)[4] and Crum-Brown (1875)[3] by their animal experiments on the labyrinth directed attention to importance of the otolith as a gravity receptor organ. About 50 years later the subject of function of the otoliths was again made current by the valuable animal work of Magnus and De Kleyn (1924)[5]. Hence when Barany (1921)[6] first became aware of the strange and dramatic vertigo which occurs in certain head positions he was led to describe the condition as otolith nystagmus. He described two distinct conditions: One attributed to otolith disorder (Barany's first type); the others attributed to a lesion within the central nervous system (his second type). According to Dix and Hallpike, Barany had not a great deal to say about either of these conditions, but what he did say was very much to the point, and in due course we shall return to his own words upon the subject.

Since then many papers have been written on the subject in many languages. Nylen's (1931)[7] clinical and experimental studies are well known, in particular his monograph on positional nystagmus occurring in intracranial tumours. In a survey of the subject in 1950 Nylen[7] gives a bibliography of no less than 297 papers by 192 authors. The clinical importance of positional nystagmus lies in the fact that its appearance is proof of an organic disorder of the vestibular mechanism either in the peripheral or in the central organ. Moreover, the characteristics of the nystagmus may be help in localizing the disturbance.

Positional nystagmus is considered to be one of the most objective and easily demonstrated signs of disturbance in the vestibular system. A great advantage in the search for this nystagmus is that the equipment used in the tests is the simplest possible.

DEFINITIONS

According to Nylen, positional nystagmus is a spontaneous nystagmus, the arising of which is influenced by the position of the head. Only that nystagmus, which changes its direction in certain head positions is

recongnized by them as true positional nystagmus. From the clinical point of view it is important to differentiate between spontaneous and positional nystagmus. Spontaneous nystagmus is one which is not influenced by the position of the head, but which is constantly alike in all head positions, while positional nystagmus is one way or another is clearly altered when the head adopts in another posture. As to the differentiation from the other type of nystagmus, Lindsay considered that positional nystagmus is differentiated from the nystagmus which in some cases may by activated by the head shaking test. The head shaking test and the postural tests have no definite relationship. The former provides an abnormal strong stimulus which tends to activate a latent spontaneous nystagmus. on the other hand the postural tests consist of physiological movements to bring the head and body into a series of positions and do not tend to activate a latent nystagmus. It should be remembered that positional nystagmus is also differentiated from the positive fistula reaction as well as the nystagmus which in rare cases may be produced by applying pressure over the vessels in the carotid sheath.

CLASSIFICATION

Several classification of positional nystagmus proposed are known by author's names, those of Nylen (1950)[7] and Lindsay (1945)[8] are the best known. However, Seiferth (1937), Ruttin (1936), Frenzel (1938), Aubry (1954) and Aschan (1956) have also proposed the classification of it.

Of these classifications there have been some agreement as to three main types which appears to have some significance in differetiating a central from a peripheral lesion, even if the nomenclature varies.

The above mentioned authors base their classification on the direction and duration of the nystagmus.

Type I. Direction-changing positional nystagmus. (Nylen's type I, Seiferth's type l, Ruttin's type l, Lindsay's type l, Frenzel's true positional nystagmus).

The direction is changed in certain positions of the head, usually being reversed in the opposite positions in this type. It may be horizontal in one position and may be vertical, oblique or rotatory in some other position.

Type II. Direction-fixed positional nystagmus. (Nylen's type II, Frenzel's spontaneous nystagmus released by position stimulus).

In this type the nystagmus may appear in the same direction when it arises. If it is observed in all head positions it is clearly altered in appearance and above all in strength in a certain or in certain positions.

Type III. Irregular positional nystagmus.

This kind of nystagmus is characterized by variations in its behaviour. It is thus sometimes direction-changing, sometimes direction-fixed, and sometimes changing its direction in the same head position.

Positional nystagmus may be persistent (of unlimited duration in a given position), or transitory (of brief duration, lasting anywhere from 10 to 60 seconds). Aschan et al (1956) have described the following classification:

Type I. Nystagmus persistent, direction-changing.

Type II. Nystagmus persistent, direction-fixed.

Type III. All varieties of transitory positional nystagmus.

The most frequently observed type of positional nystagmus in Type III is benign paroxymal type described by Dix and Hallpike (1952).

METHOD OF EXAMINATION FOR POSITIONAL NYSTAGMUS

Although this tests should theoretically be done with the condition of the head and body in the same relationship to avoid neck reflexes, in the human subjects the neck reflexes have a negligible effect on eye movements and do not complicate the tests. Grahe (1927), Fromm & Nylen (1935) have used special tables for positional tests. For clinical purpose, it seems to be adequate by using an ordinary examiantion table. In general observations of the eyes for the nystagmus may be done with the use of +20 diopter lense to eliminate fixation. It is available to utilize electronystagmography for recording the positional nystagmus. A routine sequence in testing the various positions is advisable, and should be adhered to if possible.

According to Lindsay[9] the order in which the tests have been found to be convenient is as follows: 1. Sitting on the table in erect posture. 2. From the erect to supine. 3. From supine to right lateral. 4. From right lateral to supine. 5. From supine to left lateral. 6. From left lateral to supine. 7. Head extended over the end of the table. 8. Supine position again. 9. From supine to erect. 10. Feet over the side of the table and stooping forward to the head-hanging position, and 11. From stooping over to the erect position. In order to avoid the technical error during head movements it should be kept in mind that change of position should take place very slowly. Usually position can be changed 90° in about 5 seconds.

RESULTS OF THE PRESENT STUDY

We come, now, to present results of our study, made possible through the courtesy of our colleagues in the Otological Department of Yamaguchi

University, of a consective series of 213 cases presenting positional nystagmus. In our cases the vestibular signs and symptoms have been observed and particular attention has been directed towards the analysis of characteristics of positional nystagmus. These we have examined in the course of the last few years.

Age and sex distribution are tablated in Table 1. The great majority is included in the age group 20 to 50 without preferance for sex.

In 213 cases of our present study we found twenty five cases (21 per cent) of direction-changing positional nystagmus (Type I), one hundred fifty eight cases (74 per cent) of direction-fixed (Type II) and ten cases (5 per cent) of irregular (Type III). It will be seen from Table 2 that the great majority in our cases is included in the direction-fixed type of positional nystagmus.

Table 3, 4 and 5 show in our cases localizations of the lesions or names of the diseases in each type of positional nystagmus.

Table 1.

Age Distribution

Age (Years)	Under 20	20-29	30-39	40-49	50-59	over 60	Total
Number	10	45	80	47	28	3	213

Sex Distribution

Male 115
Female 107 Total 213

Table 2. Positional Nystagmus

Type Ⅰ. Direction-changing	45	21%
Type Ⅱ. Direction-fixed	158	74%
Type Ⅲ. Irregular	10	5%
Total	213	100%

Table 3. Direction-fixed Positional Nystagmus Type II

Middle ear disease	22	Facial paresis	4
Meniere's disease	22	Hormonal lesion	3
Vestibular neuritis	20	Spinal lesion	2
Labyrinthine fistula	15	Narcotic intoxic.	1
Sudden deafness	5	Strabismus	1
Head injury	19	Sympathetic N. L.	1
Central nervous L.	5	Unkown	13
Cerebro-vascular D.	29		
		Total	158

Table 4. Direction-changing Positional Nystagmus Type I

Central nervous L	13	Head injury	3
Cerebro-vascular D.	12	Middle ear D.	2
Sudden deafness	5	Labyrinthine L.	1
		Total	45

Table 5. Irregular Positional Nystagmus Type III

Labyrinthine lesion	3	Cerebro-vascular D.	1
Head injury	3	Central nervous L.	1
Middle ear disease	2		

COMMENT

In comment we propose now to consider in detail some important clinical aspects of positional nystagmus.

1. The clinical importance of positional nystagmus.

In general we are able to find out no positional nystagmus in normal subjects, so that presence of positional nystagmus in any type, beyond any doubt, make it apparent that some disturbance is present either peripheral or central in the vestibular system. This vestibular system includes the organ of equilibrium such as the utriculus, sacculus and semicircular canals in the inner ear, nervus vestibularis and centres in the brain. Although there is the doubtful opinion that positional nystagmus only appears in central lesions, it is greatly important to point out that this nystagmus can appear in diseases in regions widely disturbing the vestibular system.

According to Nylen[7] positional nystagmus may be present, where the inner ear is concerned in, e. g. inflammation, haemorrhage, thrombosis, circulatory and secretory disturbances, degeneration and toxic condition; regarding nervus vestibularis - in neuritis, tumours, vascular anomalies, degeneration, atrophy, etc.; concerning the brain - in encephalitis, tumours, abscess, increased intracranial pressure, vascular disturbances, cranial trauma, haemorrhage and toxic conditions. Nylen concluded in his experience that direction-changing and irregular positional nystagmus speak first for a central disturbance, while direction-fixed positional nystagmus may appear in labyrinthine and retrolabyrinthine disturbances as well as in central lesions.

2. Positional nystagmus in peripheral lesions.

(1) Middle ear diseases.

It is agreed by many otologists that middle ear disease of non-suppurative or inflammatory types is occasionally accompanied by vertigo and may be present positional nystagmus. In a series of 571 cases of acute middle ear infection in hospitalized and out-patients, Blomqvist reported a nystagmus affected by position in 26 of 31 cases with pathological nystagmus. In the great majority of these cases, positional nystagmus was direction-fixed towards the affected side, while 3 cases were direction-changing, and only one case was direction-fixed towards the normal side from the beginning of the disease. In 5 cases the direction of the nystagmus was changed during the course of the disease from the affected to normal side.

In 26 cases of middle ear disease in our present study we found 22 cases with direction-fixed positional nystagmus, 2 cases with direction-changing and 2 cases with irregular. In 22 cases with direction-fixed positional nystagmus we found 18 cases of direction towards the normal side and 4 cases of direction towards the affected side of which direction changed from the affected to the normal side during the course of disease with subsided vertigo. So that all our cases with direction-fixed nystagmus of our study showed the direction towards the normal side at the end of course of the disease.

(2) Labyrinthine lesions.

a) Labyrinthine fistula.

Patients with labyrinthine fistula complain of recurring onsets of vertigo and at times of nausea which are able to be triggered by turning the head and body quickly, and lying down in acute stage. A positional nystagmus towards the affected side may be present in this stage, while in chronic stage this nystagmus towards the normal side may or may not be

appeared.

In 15 cases with labyrinthine fistula of our present study we found all of the cases with direction-fixed positional nystagmus towards the normal side and no case with direction-changing or irregular positional nystagmus. In these cases, in addition, labyrinthine fistula of the semicircular canals was identified by surgical procedures.

b) Vestibular neuritis.

According to Stahle (1966), the patients examined during an early stage of vestibular neuritis showed usually positional nystagmus towards the normal side in unilateral cases.

In 20 cases with vestibular neuritis of our present study we found 16 cases with direction-fixed positional nystagmus, 3 cases with irregular and one case with direction-changing. In eleven of 16 cases with direction-fixed positional nystagmus, the direction of the nystagmus was towards the normal side from the beginning of the disease, while 5 cases were the direction towards the affected side. In 4 cases the direction of the nystagmus was hanged during the course of the disease from affected to normal side.

c) Meniere's disease.

In Meniere's disease or labyrinthine hydrops, during an onset when a spontaneous nystagmus is present in all positions, the intensity may be influenced by position. The inclusion of such cases under positional nystagmus materially increases the incidence in that disease. Lindsay emphasized that in contrast to Meniere's disease a positional nystagmus has been observed so frequently in vestibular neuritis that it may be considered as a characteristic.

In 22 cases with Meniere's disease of our present study, we found 18 cases of direction-fixed positional nystagmus towards the normal side and 4 cases of the same type of nystagmus towards the affected side which are considered to be hyperexcited in stage.

d) Sudden deafness.

Sudden deafness has been described with increasing frequency in recent years. This disease presents with history of an apoplectic onset of hearing loss often associated with vertigo and tinnitus. In the majority of cases only one ear is affected. Usually these symptoms in all periods from the third decade of life onward, without clear cut evidence of associated disease, has left the etiology undecided. As to the etiology of this disease, many authors thought three likely possibilities: occulusion of the internal auditory artery by spasmus; subclinical mumpus; and a single episode of

Meniere's disease. Lindsay stated that the positional nystagmus observed in sudden deafness has been of the direction-changing type, lasting usually less than one minute, but reversal of the nystagmus in opposite positions of the head has not been observed.

In 10 cases with sudden deafness of our study, we found 5 cases of direction-fixed positional nystagmus and the other 5 cases of direction-changing type. In two of 5 cases of direction-fixed type, the direction of nystagmus was towards the affected side during the full course of the disease, while in remaining three cases the direction was towards the normal side.

3. Positional nystagmus in diseases with uncertain localization.

In this series of cases the vestibular disturbances may or may not be accompanied by cochlear disturbances such as hearing loss or tinnitus and the clinical examination shows no exact signs of disease of the central nervous system. Clinically we, otologists, consider to include the following diseases in this group: intoxications from drugs or infections, hypertension hypotension, vasomotor instability, menopause and head injury. In the above mentioned diseases there may be no evidence as to definite etiology up to date. It is generally accepted that streptomycin or kanamycin may affect the vestibular nuclei primarily, others such as quinine and salicylates disturb either the vestibular or cochlear region.

According to Lindsay in hepertension positional nystagmus may be present at the onset and in hypotension, vasomotor instability and menopause the positional nystagmus usually of direction-changing type appear in the majority of cases. Head injury is recognized as a common cause of vertigo. In this lesion the positional nystagmus in direction-changing in type is most frequent and suggests a disturbance in the vicinity of the vestibular nulcei.

In 25 cases with head injury of our present study we found 19 cases of direction-fixed type, cases of direction-changing type and 3 cases of irregular type. In eighteen of 25 cases of direction-fixed type the direction of the nystagmus was towards the normal side and in the remaining seven the direction was towards the affected side.

As seen the above mentioned data in our present study, the majority of cases with head injury appeared the direction-fixed type of positional nystagmus, while according to Seiferth, Nylen and Lindsay in this disease direction changing or irregular types of positional nystagmus are very common. It should be kept in mind that in our present study all of cases with head injury were not severe in condition without skull fracture.

4. Positional nystagmus in central lesions.

In cases of brain tumour positional nystagmus is a common symptom. Nylen in a series of 673 varified cases of brain tumour demonstrated positional nystagmus in 279 of the 360 cases with nystagmus. In 50 per cent the positional was direction-fixed, in 30 per cent direction-changing and in 20 per cent the nystagmus was spontaneous.

In subtentorial tumours positional nystagmus of all types was found in 69 per cent of 240 cases, while in supratentorial tumours the incidence was 26 per cent of 433 cases. Nylen also observed that in tumours of the posterior fossa the positional nystagmus most frequently occurred in the side positions or many positions while in supratentorial lesions it was most frequent in the hanging head position.

In 19 cases with central nervous lesions of our present study we found five of direction-fixed type, thirteen of direction-changing and one of irregular of positional nystagmus.

In 42 cases with cerebro-vascular disease of our present study we found twenty nine of direction-fixed type, twelve of direction-changing type and one of irregular type of positional nystagmus.

5) Pathogenesis of positional nystagmus.

We come, finally, to discuss pathogenesis of positional nystagmus, which seems to be of importance making diagnosis. Appearance of positional nystagmus can be explained by assumption of altered tonus in the vestibular systems. From this point of view, in direction-changing or irregular positional nystagmus the tonus-preponderance in the vestibular system of both sides asserts itself by turns, but only in certain positions of the head, while direction-fixed positional nystagmus appears as a result of tonus preponderance in the vestibular system of one side when the patient adopts certain head positions.

a) In peripheral lesions.

In labyrinthine disturbances it is conceivable that positional nystagmus arises through deficient interplay of the otoliths and the cupulae (Lund), corresponding to a peripheral direction preponderance in agreement with the theories of the significance of the utriculus-otolith in abnormal caloric nystagmus of Cawthorne, Hallpike and Jongkees. Nylen adheres to the theories held by Kobrak, M.H.Fischer, Frenzel and Ohm who maintain that the labyrinthine changes cause a central direction preponderance, because the pathological peripheral stimuli lead to and charge the vestibular centres to a greater or lesser extent. Frenzel remarks that all types of nystagmus arise centrally.

According to Nylen, the following nystagmus appear when an acute inflammatory process extends from the middle ear into the inner ear:

1st stage; A direction-fixed positional nystagmus towards the affected side arises because the interplay of the otoliths and the cupulae on the affected side are disturbed by colateral hyperaemia, and colateral edema beginning in the cysterna perilymphatica.

2nd stage; Spontaneous nystagmus to the affected side because the entire vestibular organ is irritated equally by progression of the pathological changes just mentioned tonus preponderance in the vestibular centres of the affected side.

3rd stage; Direction-changing positional nystagmus because the interplay of the vestibular organs is again disturbed, probably because of more pronounced changes in the interior of the utriculus, while the cupulae still function. The pathological changes in the perilymphatic and endolymphatic rooms have advanced to a serous process.

4 th stage; Direction-fixed positional nystagmus towards the normal side, presupposing further progress of the serous inflammation, whereby the utriculus otolith is quite paralysed and even the cupulae in the affected side begin to be pue out of action.

5 th stage; Spontaneous nystagmus towards the normal side, the cause of which is that the otoliths as well as the cupulae have ceased to function. The pathological process is on the way to becoming purulent, or is already so. It is now the labyrinth of the normal side which is responsible for the spontaneous nystagmus towards the normal side and for the central tonus preponderance which has hereby arisen on the normal side.

b) In central lesions.

It is agreed by the great majority of authors (Barany, Kobrak, de-Klen, M.H. Fischer, Spiegel, Brunner and Nylen) that if a pathological condition is present in different parts of the vestibular systems in the brain, stimuli flow from both the normal labyrinths to the vestibular centres. The balanced inter-relation of these centres is disturbed by the affected changes there. Under these conditions a central tonus difference, etc. arises, which can also be produced by a cessation of the inhibition mechanisms if the disease process has affected parts of the supra-vestibular system in the cerebellum and cerebrum were such inhibition centres are thought to be situated.

According to Nylen the primary cause of the vestibular tonus differences which arise in the vestibular system in labyrinthine, retro-labyrinthine and central affections must be sought in the first place in

abnormal metabolism in the nerve cells of the vestibular sense organ, the ganglion cells and in the nerve cells in the various centres.

SUMMARY

In a consecutive series of 213 cases presenting positional nystagmus, classification of the nystagmus type, localizations of the lesions and pathogenesis of positional nystagmus were discussed.

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