

VESTIBOLOGY

The clinical significance of direction-fixed mono-positional apogeotropic horizontal nystagmus

Il significato clinico del nistagmo monoposizionale orizzontale apogeotropo

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SUMMARY

Objective. A mono-positional persistent, direction-fixed apogeotropic nystagmus (MPosApoNy) is very challenging for the neuro-otologist. MPosApoNy can be found in patients suffering from a partially compensated acute unilateral vestibulopathy; with a normal caloric test, one can speculate the presence of “trapped” otolithic debris located close to the ampulla of the horizontal semicircular canal.

Methods. Among 957 patients suffering from vertigo and dizziness, we selected 53 cases of MPosApoNy.

Results. In 28 patients, caloric test showed a canal paresis on the same side of the MPosApoNy. In the remaining 25 cases, MPosApoNy was the only clinical finding. We hypothesized the presence of horizontal canal lithiasis and patients were treated with a Gufoni manoeuvre, followed by a forced prolonged position.

Conclusions. Performing bedside examination in a patient suffering from vertigo, the presence of MPosApoNy may be due: a) facilitation of a subclinical nystagmus due to the mechanism of apogeotropic reinforcement; b) horizontal canal lithiasis with ‘trapped’ otoliths close to the ampulla. The disappearance of MPosApoNy following a repositioning manoeuvre or conversion in a typical form of canalolithiasis may represent the best method to confirm this hypothesis.

KEY WORDS: benign paroxysmal positional vertigo, horizontal semicircular canal, apogeotropic nystagmus, positional nystagmus, canalolithiasis

RIASSUNTO

Obiettivo. Il riscontro di un nistagmo monoposizionale orizzontale apogeotropo (MPosApoNy) rappresenta una sfida diagnostica per il vestibologo. Tale nistagmo può indicare, in presenza di un deficit al test calorico, una labirintopatia periferica non compensata. In assenza di tale reperto, il MPosApoNy potrebbe essere riconducibile ad una forma atipica di litiasi del canale semicircolare orizzontale (CSL).

Metodi. Su 957 pazienti con vertigini, ne abbiamo selezionati 53 che presentavano MPosApoNy. **Risultati.** In 28 pazienti il test calorico evidenziò un deficit ipsilaterale al MPosApoNy. Nei rimanenti 25 in cui il test calorico risultò nella norma, si ipotizzò una litiasi del CSL con otoliti intrappolati in prossimità dell’ampolla. Questi pazienti vennero trattati con manovra di Gufoni e successiva posizione coatta.

Conclusioni. Il riscontro di un MPosApoNy può riconoscere una duplice causa. Esso può essere l’espressione di un nistagmo subclinico esito di labirintopatia acuta monolaterale. Tuttavia, la presenza di otoliti intrappolati in stretta vicinanza dell’ampolla ne può spiegare la genesi. In quest’ultimo caso, le manovre liberatorie, attraverso la risoluzione del MPosApoNy o per la conversione in una forma tipica di canalolithiasis, possono confermarne l’origine litiasica.

PAROLE CHIAVE: vertigine parossistica posizionale benigna, canale semicircolare orizzontale, nistagmo apogeotropo, nistagmo posizionale, canalolithiasis

Introduction

Positional nystagmus is defined as a nystagmus triggered by and occurring after a change in head position with respect to gravity ¹. This nystagmus can

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be transient (as in benign paroxysmal positional vertigo) or persistent: in the latter case the duration is long (generally > 1 minute) and the intensity is constant or slowly decreases over time. The direction of persistent positional nystagmus can be the same in both ear-down positions (ipsiversive or unidirectional, fixed) or can change (contraversive or bidirectional) ². Occasionally, a persistent direction-fixed (ipsiversive) positional nystagmus can manifest only in one ear-down position. This behaviour can be observed in patients suffering from acute unilateral vestibulopathy in whom the spontaneous nystagmus continues supine, but its intensity increases when the patient lies on the affected side (affected ear down, homolateral excitation) and decreases on the opposite side (unaffected ear up, contralateral inhibition) ^{3,4}. As the vestibular tone imbalance causing spontaneous nystagmus progressively decreases (by means of the recovery of vestibular function or central vestibular compensation) until it disappears in sitting position, when performing the positional test the nystagmus can appear only when the patient's head is rotated to the affected side when supine (apogeotropic reinforcement). This nystagmus will be positional, persistent and present only on one side (mono-positional and apogeotropic) ⁵. Persistent positional nystagmus has also been described (mostly bidirectional) in some central vestibular diseases such as vestibular migraine ⁶, more often in association with other signs of central nervous system involvement. In some cases, a persistent ipsiversive positional nystagmus (the nystagmus does not reverse its direction with the supine roll test) may be attributed to an atypical form of lateral canal benign paroxysmal positional vertigo ⁷. In this paper, we retrospectively describe the characteristics of a population of patients suffering from vertigo showing a persistent horizontal apogeotropic mono-positional nystagmus.

Materials and methods

Among 957 patients admitted to our tertiary referral centre suffering from vertigo and dizziness in the period from January 2015 to December 2019, we selected 53 patients showing a mono-positional apogeotropic horizontal nystagmus. The 53 patients came to our observation because of the onset of acute vertigo exacerbated or occurring after a change of head position in space relative to gravity. All patients, after careful assessment of clinical history, underwent a complete vestibular examination, consisting of the search for ocular alignment, spontaneous nystagmus, horizontal and vertical gaze-holding nystagmus, head shaking nystagmus, horizontal and vertical saccades, horizontal and vertical smooth pursuit and head impulse test. Positional

manoeuvres included lying down from sitting, head turning to either side while supine (supine roll test), straight-head hanging and Dix-Hallpike test. Caloric test was performed according to a modified Fitzgerald–Hallpike technique. The responses were recorded using an infrared eye-tracking system (GN Otometrics, Taastrup, Denmark); canal paresis (CP) was considered significant if > 25%. All patients underwent MRI investigation to rule out an involvement of the central nervous system.

Results

All 53 patients (32 males and 21 females age ranging from 20 to 84 years of age, mean 58 years) showed a persistent positional horizontal apogeotropic nystagmus in a single position of the head (left or right side) during the supine roll test and Dix-Hallpike test. In 21 patients there was an apogeotropic nystagmus on the left side, and in 32 patients there was an apogeotropic nystagmus in the right side. In 28 of the 53 patients, caloric test showed a canal paresis on the same side of the apogeotropic persistent positional nystagmus; in all patients with canal paresis there was a congruence between the side of reduced caloric response and the side in which the positional nystagmus appeared. The head shaking test was positive with a peripheral pattern in 26 of 28 patients with canal paresis and in 4 patients with normal caloric test. In the remaining 25 cases, apogeotropic mono-positional nystagmus was the only clinical and instrumental finding. In these cases, we hypothesised the presence of horizontal canal lithiasis and patients were treated with a Gufoni manoeuvre ⁸, followed by a forced prolonged position ⁹.

After one session of treatment, 9 patients showed disappearance of the positional nystagmus and in 5 cases a conversion in a typical form of horizontal canal benign paroxysmal positional vertigo (HC-BPPV, 4 apogeotropic and 1 geotropic) occurred. In one case a canal switch towards an involvement of the posterior semicircular canal (PC-BPPV) was evident. Five patients showed the disappearance of the nystagmus after the second session of treatment and in two cases a conversion in an apogeotropic form of HC-BPPV was registered. The remaining 3 patients were instructed to perform a series of home exercises consisting in four daily sessions of head shaking test and barbecue manoeuvre. At one-month follow-up, disappearance of the monopositional persistence was evident (Fig. 1).

Brain MRI showed normal results in all patients.

Discussion

We speculate that the presence of an apogeotropic posi-

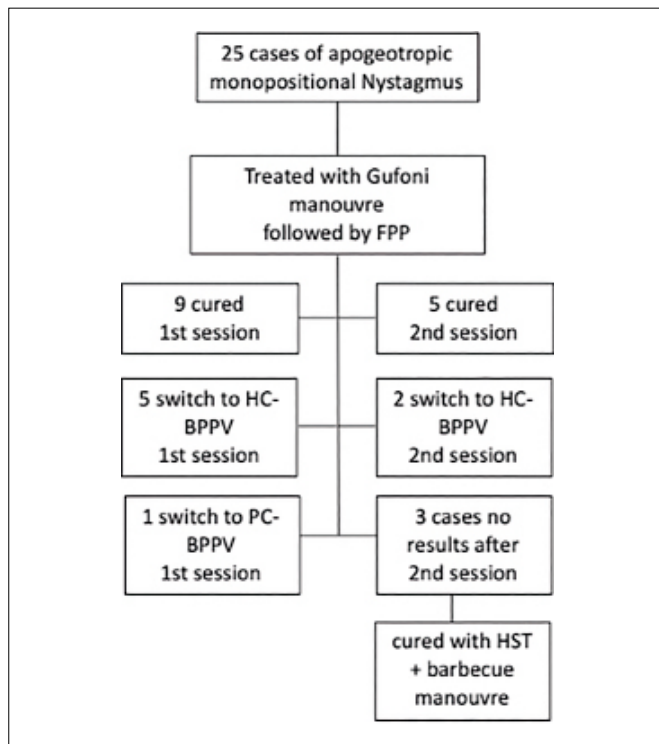


Figure 1. Summary of the results obtained with treatment in the 25 patients with monopositional apogeotropic horizontal nystagmus.

tional nystagmus in only one position may be due to two pathological conditions:

1. an uncompensated unilateral vestibular loss;
2. an atypical form of horizontal semicircular canal lithiasis.

An apogeotropic mono-positional horizontal nystagmus can be associated with a canal paresis, and in this case it can be considered as the result of progressive improvement of the imbalance between the two sides after unilateral vestibular damage. A reduction of otolithic function on the affected side might reinforce a spontaneous nystagmus when the patient lies on the affected side³. In patients with unilateral complete peripheral deafferentation, it is possible to observe an increase of the intensity of the spontaneous nystagmus when positioned on the affected side (the nystagmus is apogeotropic) and a reduction of its intensity when the patient lies on the healthy side (the nystagmus is geotropic)¹⁰. This phenomenon is related to asymmetric macular activation due to unilateral macular damage. As vestibular compensation or spontaneous recovery progress, the vestibular tone imbalance causing spontaneous nystagmus progressively decreases; consequently, the nystagmus can be seen only by moving the head (i.e., performing a Head Shaking Test) or performing the positional test. In the

latter, an apogeotropic mono-positional horizontal nystagmus can be detected as the only expression of the imbalance of the two vestibular hemi-systems. In other terms, the position of the head reinforces the nystagmus, which becomes clinically evident. This condition can be easily confirmed by caloric tests, from which we expect a canal paresis, i.e. expression of the reduced function of one horizontal semicircular canal. Indeed, the results of the head shaking test were congruent (beating to the unaffected side) with the lesioned side in the majority of the 28 patients showing a canal paresis.

The 25 cases in which the apogeotropic mono-positional horizontal nystagmus was not associated with canal paresis remain to be interpreted. An apogeotropic mono-positional nystagmus in the absence of signs of unilateral vestibular loss could be hypothetically considered as the consequence of a lateral canal lithiasis in which otolithic debris remain ‘trapped’ within the ampulla (Fig. 2). We can speculate that the dislocated otoliths are located close to the ampulla of the lateral semicircular canal (in our example on the right side) and are blocked in a cluster larger than the diameter of the channel (narrow part) and smaller than the diameter of the ampulla, with the result that the debris can move within the ampulla but cannot fall into the canal. If the patient assumes the right-side position when lying down, the debris must stop at the narrowest part of the canal. The movement of the debris inside the ampulla is not able to generate a nystagmus¹¹; consequently, in the right side position no nystagmus can be elicited. When the patient turns the head

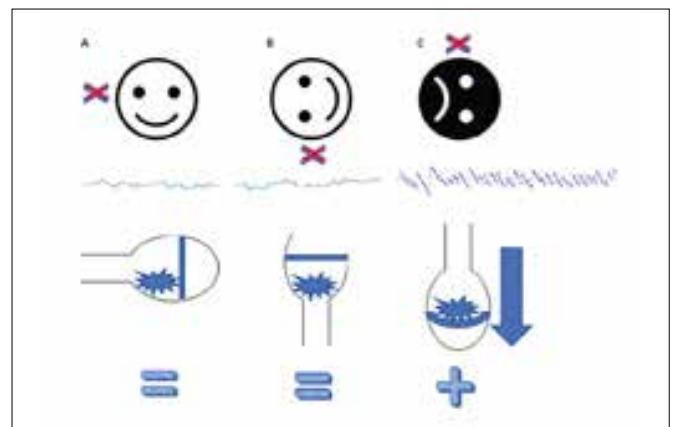


Figure 2. The apogeotropic monopositional nystagmus from ‘trapped’ otolithic block. (A) The patient is in sitting position: the otolithic block is close to the right ampulla. (B) When the patient lies supine with the head rotated to the right, the block does not provoke any cupular movement and no nystagmus is evident (the otolithic debris does not enter in the canal because of its dimension (=)). (C) When the patient lies supine with the head rotated to the left, the movement of debris creates a persistent ampullopetal displacement of the cupula and an apogeotropic nystagmus appears (+).

to left side, the debris could induce an ampullopetal deflection of the cupula which provokes an excitatory nystagmus directed to the right, and is apogeotropic because the patient is positioned on the left side. The result is an apogeotropic nystagmus evoked positioning the patient on the left side with no nystagmus in the opposite position. An origin of the above-described positional nystagmus from a condition of heavy/light cupula¹² seems to be less probable: usually this condition should cause a positional apogeotropic nystagmus when turning the head to the right side and to the left side (bidirectional) when the patient lies in supine position. The presence of otolithic deposits in atypical locations has already been described, namely the finding of a paroxysmal nystagmus with fixed direction, apogeotropic on the pathological side, and geotropic on the healthy side⁷. The positional nystagmus recorded in our series of patients differ from the latter because it appears in only one position of the head (right or left) when the patient lies supine. Although a central origin of the apogeotropic positional nystagmus (monodirectional or bidirectional) cannot be excluded¹³, our series of patients showed a normal brain MRI. Our observations seem to suggest the possibility of a very particular and not so common type of lithiasis of the semicircular canal in which the debris are “trapped” close to the ampulla, and unable to move into the narrow tract of the canal. This situation would be associated with a normal caloric test, but a reduction of caloric response cannot be *ex ante* excluded^{14,15}.

Based on this hypothesis, we tried to treat these patients using a combination of repositioning manoeuvres. The disappearance of the apogeotropic mono-positional nystagmus after performing one or two sessions of treatment, or its conversion in a feature compatible with a typical involvement of the horizontal or posterior semicircular canal lithiasis, could represent the confirmation of our hypothesis and could be the best evidence for a differential diagnosis^{16,17}. In some cases, the inefficacy of the repositioning manoeuvre could be due to debris that has fallen into the short arm of the semicircular canal, as previously described¹⁸. Recently, a high percentage (38.7%) of patients suffering from Menière’s disease showed a monopositional apogeotropic nystagmus when turning the head to the affected side. This observation could be related to the comorbidity between Menière’s disease and vestibular migraine, conditions commonly characterised by the presence of a positional nystagmus¹⁹. However, in our series we excluded patients with clinical and instrumental findings compatible with a definite diagnosis of Menière’s disease.

Based on the above-described observations, in a patient complaining of vertigo and showing a monopositional apogeotropic nystagmus, the diagnostic strategy could be set up as follows:

- in the presence of a monopositional apogeotropic nystagmus, it is always advisable to complete the examination with caloric tests. In the case of congruent dysreflexia, it is reasonable to interpret the nystagmus evoked as a facilitated subclinical nystagmus. It is to be expected that, over time, the nystagmus tends to disappear along with the improvement of the compensation and normalization of the directional preponderance;
- if the caloric test shows no canal paresis, an atypical form of HC-BPPV can be suspected. The hypothesis is that the debris is in the horizontal canal on the side where no nystagmus is evoked. In case of modification of the nystagmus after the manoeuvre (disappearance or conversion into a HC-BPPV or PC-BPPV), the diagnosis moves towards a form of ‘trapped’ ampullar lateral canalolithiasis. We suggest performing the repositioning manoeuvres even in the case of mild asymmetry of the caloric response, especially if the patient reported a clinical history predominantly characterised by positional vertigo. It is well known that the presence of a bulky otoconial mass may be associated with mild hypofunction of the HC.

Finally, we must take in consideration that positional nystagmus (especially horizontal direction-changing apogeotropic or geotropic) may occur in up to 88% of healthy subjects²⁰. Moreover, central nervous system involvement could produce a positional nystagmus appearing mostly as downbeat nystagmus while prone or supine, or apogeotropic or geotropic horizontal nystagmus when the head is turned to either side while supine¹³.

Our study has several limitations: firstly, not all patients were investigated using the video Head Impulse test, which could provide more information about the functionality of the two labyrinths at more physiological stimuli with respect to the caloric test; secondly, we do not have a control group as in some normal subjects a positional nystagmus can be detected. Furthermore, we assumed that modulation of the spontaneous nystagmus due to a unilateral vestibular loss while performing the positional tests is related to asymmetric macular activation due to the unilateral macular damage; to confirm this hypothesis, a study of the utricular and saccular function would be needed. Finally, we did not perform cervical and ocular Vemps in all patients of our cohort. This could have prevented the discovery, although very rare, of some type of positional nystagmus of macular origin²¹.

Conclusions

When performing a bedside examination in a patient suffering from acute vertigo, the finding of apogeotropic mo-

nopositional nystagmus can be difficult to interpret. We suggest two possible diagnostic keys in order of clinical frequency, as regards the differential diagnosis:

1. facilitation of a subclinical nystagmus due to the mechanism of apogeotropic reinforcement. In this case, it is also possible to find a head shaking nystagmus directed in the same direction of the positional nystagmus, congruent with the reduced activity of the HC as detected with the caloric test;
2. HC-BPPV with 'trapped' otoliths close to the ampulla. The only method to confirm this hypothesis consists in performing a liberatory manoeuvre, assuming the side where the nystagmus does not appear as the affected side. The disappearance of this nystagmus or a conversion in a more common form of BPPV may represent the best method to confirm this hypothesis.

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Conflict of interest statement

The authors declare no conflict of interest.

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Authors' contributions

PLEASE COMPLETE

MG: study concept and design, acquisition of data, analysis and interpretation of data, study supervision.

APC: study concept and design, analysis, and interpretation of data, drafting of the manuscript, study supervision.

Ethical consideration

Ethical review and approval by the local Institutional Board (Comitato Etico Azienda Ospedaliero-Universitaria Pisana, Pisa, Italy) were waived for this study. Due to its retrospective nature, it was not considered as part of a research project. Furthermore, the study does not include new invasive or experimental procedures or diagnostic protocol; all patients underwent routinely performed tests only, according to national guidelines. Informed consent was obtained from all participants and the study was performed in accordance with the Declaration of Helsinki.

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