sciendo

DOI: 10.2478/rjr-2020-0003

Romanian Journal of Rhinology, Volume 10, No. 37, January - March 2020

LITERATURE REVIEW

Lindsay-Hemenway Syndrome: Review of the literature and case report

Alina Georgiana Anghel[1,2], Carmen Badea[3], Gabriela Musat[1,3]

[1]"Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania

[2]CESITO Centre, "Sfanta Maria" Hospital, Bucharest, Romania

[3]ENT&HNS Department, "Sfanta Maria" Hospital, Bucharest, Romania

ABSTRACT

OBJECTIVES. Reviewing the literature data related to Lindsay – Hemenway syndrome.

MATERIAL AND METHODS. We searched PubMed and Google Scholar with the key words of "Lindsay-Hemenway syndrome", "benign positional vertigo", "vestibular rehabilitation"

RESULTS. Lindsay-Hemenway syndrome is characterized by an association between vestibular neuronitis and BPPV. The specificity of the syndrome consists in the existence of an initial episode of acute vestibular neuropathy manifested by intense vertigo and nystagmus, followed in a variable time frame by episodes of posterior canal BPPV. The treatment of the syndrome consists in a combination of otolith repositioning manoeuvres and vestibular rehabilitation therapy. The physicians involved in treating patients with vestibular disorders should be aware of the existence of this syndrome in order to diagnose and treat the patients accordingly.

CONCLUSION. The Lindsay-Hemenway syndrome is a challenge for the physician. In order to establish a diagnosis, a careful investigation of clinical history and objective examination are needed. The clinician should take into consideration the presence of a sudden vertigo without deafness followed by postural nystagmus, and unilateral labyrinthine hyporeflexia or absence of reflectivity. For a successful therapeutic approach, we should be able to combine manoeuvres of repositioning for BPPV with an appropriate vestibular rehabilitation therapy in order to ensure a correct central compensation of the peripheral unilateral deficit.

KEYWORDS: Lindsay-Hemenway, repositioning manoeuvres, caloric tests, benign paroxysmal positional vertigo.

INTRODUCTION

The Lindsay-Hemenway syndrome is produced by an ischemic lesion in the territory of the anterior vestibular artery resulting in an initial episode of acute peripheral vestibular syndrome followed in a variable amount of time by recurrent episodes of benign paroxysmal positional vertigo (BPPV). The measurements of the vestibulo-ocular reflex (VOR) on the affected side show decreased or abolished response. The syndrome was described for the first time in 1956 by W.G. Hemenway and J. R. Lindsay[1].

We searched PubMed and Google Scholar with the keywords "Lindsay Hemenway syndrome", "benign positional vertigo", "vestibular rehabilitation". The search resulted in 159 titles. The literature concerning only the Lindsay-Hemenway Syndrome is very poor, although the incidence of this clinical entity is probably more important than the one reported.

EPIDEMIOLOGY

Both in the international and Romanian specialized literature, there is variable information about the incidence of the Lindsay-Hemenway syndrome. Studies conducted in Spain revealed that 16.3% of BPPV cases are related to this pathology[2].

Corresponding author: Gabriela Musat, MD, PhD, Address: ENT&HNS Department, "Sfanta Maria" Hospital, 37-39 Ion Mihalache Blvd., District 1, Bucharest, Romania

e-mail: gabimusat@yahoo.com

Received for publication: October 16, 2019 / **Accepted:** December 18, 2019

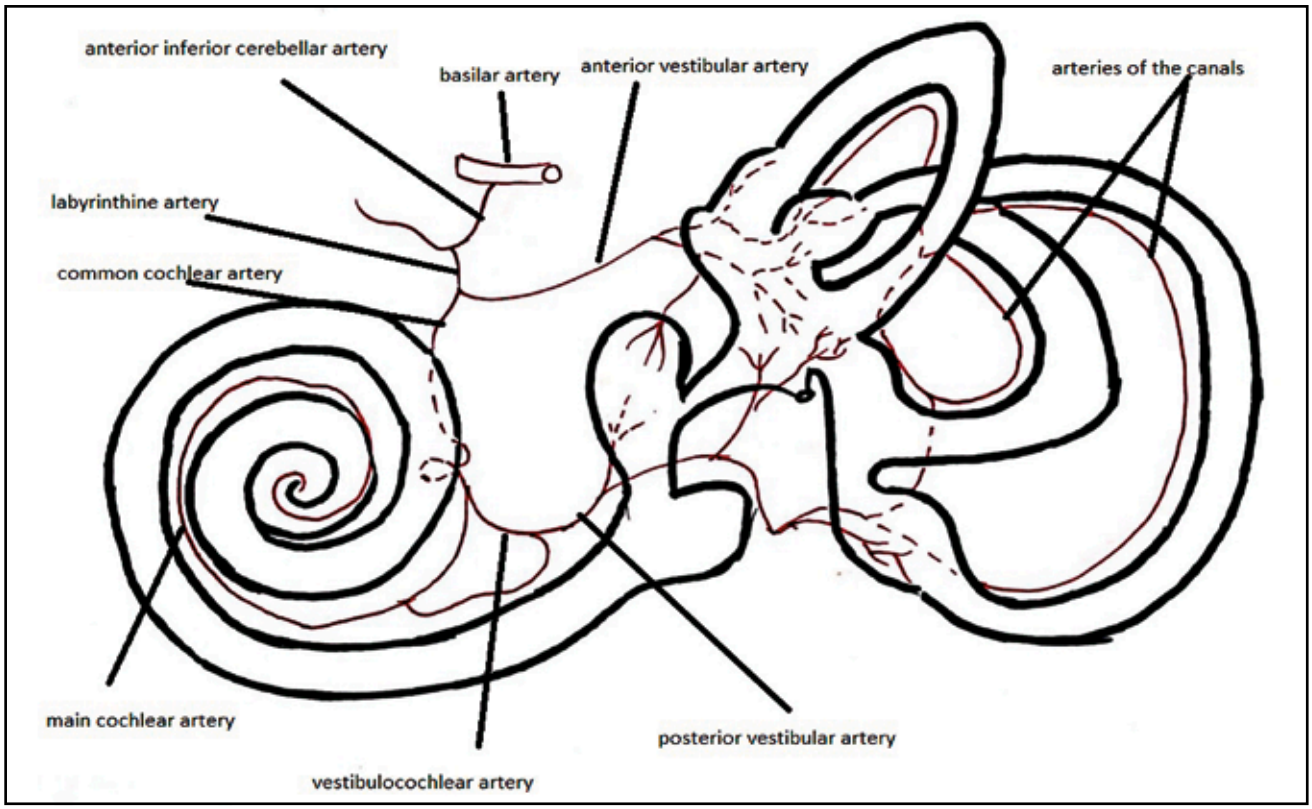

Figure 1. Vascularization of the inner ear.

It usually affects patients over 60 years, particularly those with vascular risk factors. The age category predisposed to this disease is considered to be the one between 45 and 78 years old.

The right ear is involved more frequently (66%) as opposed to the left ear (33%). According to studies, women represent 73% of cases of the pathology. Patients with Lindsay-Hemenway syndrome are predominantly from the rural area[3].

PHYSIOPATHOLOGY

The vertebrobasilar system is responsible for the vascularization of the inner ear (Figure 1). The antero-inferior cerebellar artery derives from the basilar trunk, which gives rise to the internal auditory artery. The anterior vestibular artery derives from the internal auditory artery. This artery divides into two branches inside the internal auditory canal: the common cochlear artery and the anterior vestibular artery. The anterior vestibular artery irrigates the superior and lateral semicircular canal, the utricle and a minor part of the saccule. The common cochlear artery splits at the level of the columella into the main cochlear artery and the cochlear-vestibular artery. The latter divides into the cochlear branches and posterior vestibular artery. The cochlear artery is responsible for vascularization of most of the cochlea, including the modiolus. The cochlear branches irrigate the basal turn of the cochlea. The posterior vestibular artery irrigates the saccule and the posterior semicircular canal. All the branches of these arteries are of terminal type. A consequence of this fact is the rapid structural degradation in case of interruption of blood circulation in this territory[4-6].

Lindsay-Hemenway syndrome is characterized by the interruption of blood irrigation on the territory of the anterior vestibular artery. The consequence of the ischemia is necrosis of the utricular macula, with the detachment of otoconia from the gelatinous layer of the utricular macula. Detached otoconia are displaced in the posterior semicircular canal, causing BPPV. The posterior semicircular canal is vascularized by the posterior vestibular artery. This is the reason why in case of a vascular event affecting the anterior vestibular artery, the ampulla of the posterior semicircular canal maintains its functionality and the BPPV involving the posterior semicircular canal is still possible[2].

There are two stages of the disease. The first is an acute episode of peripheral vestibular disorder which can last for more than one day and usually

responds at symptomatic treatment. The second phase is represented by the benign paroxysmal positional vertigo (BBPV) which lasts for weeks and reappears often. It is associated with no reduced to no response in caloric tests. This caloric paresis is due to the involvement of the lateral semicircular canal. It seems that the recurrence of acute vestibular neuropathy is an unlikely event whereas the incidence of BPPV after the disease is quite common[7].

DIAGNOSIS

Anamnesis plays an important role in the diagnosis of Lindsay-Hemenway syndrome. It is useful to highlight the real cause of vertigo, in this case meaning the occlusion of the vestibular artery, in order to choose the right therapy.

At the beginning, patients often describe strong sensation of dizziness triggered by the movements of the head or eyes, progressive deterioration of gait, using terms as "dizziness inside his/her head". After some time, rotatory vertigo on lying and rolling over in bed appears. The situation gets often worse in the morning when the patient stands up from bed. It is important for the clinician to give a special importance to a history of acute, intense, continuous vertigo, with a duration of hours or days, followed during weeks/months by brief episodes of vertigo associated with head movements but with no cochlear related symptoms. In some patients, the symptoms are accompanied by cervical pain and gastrointestinal discomfort. BPPV can occur immediately after the episode of vestibular neuronitis or several months later. The progressiveness of the initial symptoms can be noticed in patients with high risk of vascular thrombosis, diabetes. There is the possibility that the vestibular lesion preceding BPPV in Lindsay-Hemenway has a slowly progressive mode of installation, or that the vestibular neuronitis occurs without rotatory vertigo but only with a feeling of instability.

Otoscopy is normal in the majority of the cases and audiometry does not show alterations of the hearing function. Spontaneous nystagmus is absent at the first consultation and months after. Vestibulo-Ocular Reflex (VOR) suppression, smooth pursuit, saccades, optokinetic nystagmus (OKN) are within normal parameters. Romberg test is normal, but the patient feels unstable. Tandem gait reveals instability. Unterberger (Fukuda) stepping test shows deviation of the body toward the affected side. Dix-Hallpike and Head Impulse Test are positive on the affected side and the Supine Roll Test is negative. Caloric irrigation of the affected labyrinth shows reduced or no response[8,9].

The differential diagnosis is a difficult one because it involves two clinical entities. Cerebellum tumors, ischemic attacks, multiple sclerosis must be excluded on the basis of the identification of benign characteristics of positional vertigo. It must also be considered the presence of a positive head impulse test (HIT) and the ability of the patient to maintain the orthostatic position with eyes closed during the Romberg test[10]. AC-VEMP (air-conducted cervical vestibular evoked myogenic potentials) should be present in BPPV. In the particular situation of old people where degeneration of the saccular macula already happened, AC-VEMP are absent[11,12].

Cranial CT Angiography scan usually shows no changes in the carotid and vertebral vascular district and brainstem MRI in T2 and T1 shows no signal alteration[13].

TREATMENT

In the first phase of the disease, most patients are treated with sedatives, as they come in such a situation that does not allow the possibility to show postural nystagmus.

For the treatment of the initial episode of vestibular neuritis, clinicians usually prescribe corticosteroids, antiemetics and vestibular suppressants[14]. Some authors recommend also Acetylsalicylic acid (orally 100 mg/die), Betahistinum (orally 24mg tid). There is also the possibility to use Cinnarizine orally 75 mg/day, or Dimenhydrinate 50 mg capsules at every 4 to 6 hours in order to control nausea and vomiting[15].

When Lyndsay-Hemenway syndrome is suspected, the therapeutic Epley manoeuvre is usually performed on the affected side in order to treat the BPPV. The Epley manoeuvre consists in a sequence of positions. First of all, the patient is positioned in a sitting posture, with the legs fully extended and the head rotated 45 degrees towards the diseased side. The patient is lowered in a supine position with a 20-30 degrees neck extension. Then, the patient's head is rotated with 90 degrees toward the healthy side. The next movement is another rotation of ninety degrees toward the healthy side until the patient is looking downward at a 45-degree angle. In the end of the manoeuvre, the patient is brought up. Each position is maintained for one to two minutes. In Figure 2 it is represented this sequence of positions describing the Epley manoeuvre. The entire procedure may be repeated two more times, for a total of three times. After approximately 7 days, if the Dix-Hallpike test is still positive, we repeat the manoeuvre[16].

Figure 2. Epley manoeuvre.

In case of fail, the Semont liberatory manoeuvre can be performed. This manoeuvre turned out to be highly effective in patients presenting this disease. For a good therapeutic effect, it should be performed three consecutive times at the same session. First, with the patient seated on the examination table with her/his legs hanging, we turn the head at 45^0 towards the healthy ear. Then, the patient is asked to lie on his/her side with a neck extension of approximately 105^0 for 3 minutes towards the affected side. Patient is rapidly taken to the same position on the opposite side, turning by 195^0 face downwards resting on the contralateral ear for 3 minutes. The last step is to come back to the initial position. In Figure 3, it is represented the treatment of BPPV using the Semont manoeuvre. According to literature data, the result of the Semont manoeuvre is total cure in 90% of cases[17,18].

In some situations, we have partial cure in some cases of patients who still felt the sensation of vertigo, but did not present nystagmus at the Dix-Hallpike test. In those cases in which the BPPV is resistant to repositioning manoeuvres and the patient still experiences vertigo with or without nystagmus observable at the testing manoeuvre, the Brandt-Daroff exercises can be prescribed[19].

In order to accelerate the central compensation of the peripheral deficit, home treatment using the Cawthorne-Cooksey exercises are usually recommended[20-22]. Other recommendations are: drinking at least two litres of liquid a day; avoiding coffee in the evening, spicy food, fat meat, chocolate; following a low sodium, high potassium diet; walking as long as possible; avoiding sudden movements of the head.

CASE REPORT

We present the case of a 45-year-old woman who presented in our clinic accusing a disabling positional vertigo. The anamnesis of the patient revealed the fact that, two months prior to the presentation, the patient had had an episode of continuous vertigo associated with nausea and vomiting, with a duration of approximately one week. The vertigo she experienced had the tendency of diminishing in time until it totally disappeared in approximately one month. A few days before presentation, she started experiencing a different type of vertigo. She felt short but intense sensations of rotatory vertigo each time she moved her head.

Figure 3. Semont liberatory manoeuvre

At presentation, the ENT examination was within normal limits. Otoscopy revealed tympanic membranes with normal appearance. The audiogram we performed showed normal hearing thresholds in both ears.

At the vestibular exam, we noticed the fact that the patient did not have any spontaneous nystagmus, the Head Impulse Test was negative on the left ear and positive for the right ear. Head Shaking Test was negative, the saccade test and the slow pursuit tracking were within normal limits. The Romberg test was negative and no pathologic rotation was noticed at the Unterberger/Fukuda stepping test.

The Dix-Hallpike test was positive for the right ear revealing a posterior canal BPPV. In Figure 4, there is the videonystagmography (VNG) recording of the Dix-Hallpike right test, showing a nystagmus beating upward and to the right ear.

In the next step, we also performed a caloric test which evidenced a reduced caloric response on the right ear compared to the left one, as shown in Figure 5.

Analysing the data we have gathered from the anamnesis, the vestibular examination and the videonystagmography with caloric testing, we concluded that the diagnosis was Lindsay-Hemenway syndrome. The first episode of prolonged vertigo was the first phase of the disease, in which the vestibular anterior artery was probably partially occluded. Subsequently, two months afterwards, the episode of posterior canal BPPV was the consequence of the ischemia that fragilized the utricular macula.

We performed an Epley manoeuvre for the right ear with complete resolution of the clinical symptoms. One week afterwards, at the follow-up, the Dix-Hallpike test was negative.

Another week after this episode, the patient came once again to the hospital accusing the same positional vertigo she experienced at presentation. At

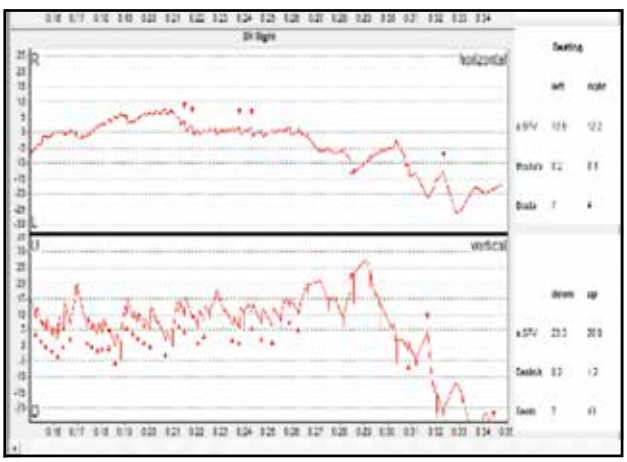

Figure 4. VNG recording of the right Dix-Hallpike testing showing an intense upbeating nystagmus with a rotatory component beating to the right undermost ear.

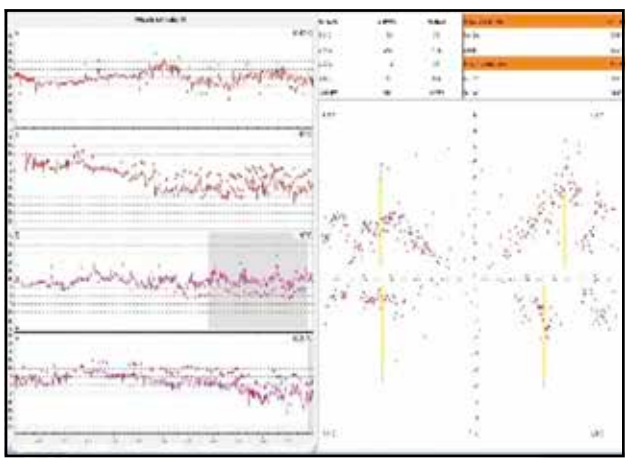

Figure 5. VNG recording of the caloric tests showing a reduced caloric response on the right ear.

the vestibular exam, we found the Dix-Hallpike test positive for the right ear. We performed the Epley manoeuvre without any effect on the symptoms of the patient. At this point, we decided to perform the Semont manoeuvre. The Dix-Hallpike manoeuvre was negative at the follow-up visit.

In this period, the patient received only neurotrophic medication to improve the process of vestibular compensation. She also received home-based rehabilitation exercises conceived as a variant of the Cawthorne-Cooksey exercises.

CONCLUSIONS

Lindsay-Hemenway syndrome is a complex clinical entity which represents a challenge for the practician. In order to establish a diagnosis, a careful investigation of the clinical history and an objective examination are needed. The clinician should take into consideration the presence of postural nystagmus, sudden vertigo without deafness and unilateral labyrinth hyporeflexia or absence of reflectivity. For a successful therapeutic approach, we should be able to combine manoeuvres of repositioning for BPPV with vestibular rehabilitation for treatment of a defective central compensation due to unilateral deficit.

Ethic approval: There was no need to take the Ethical Committee Approval.

Financial disclosure: There is no financial disclosure of the authors.

Conflict of interests: The authors report no conflicts of interest.

Contribution of authors: All authors have equally contributed to this work.

REFERENCES

1. Hemenway WG, Lindsay JR. Postural vertigo due to unilateral sudden partial loss of vestibular function. Ann Otol Rhinol Laryngol. 1956;65(3):692-706.
2. Pardal Refoyo JL, Pérez Plasencia D, Beltrán Mateos LD. Isquemia de la arteria vestibular anterior (síndrome de Lindsay–Hemenway). Revisión y comentarios Acta Otorrinolaringol Esp. 1998;49(8):599-602.
3. Gkoritsa EZ. The Lindsay-Hemenway Syndrome: two case report, review and comments. J Ear nose Throat Disord. 2016;1(1):1014.
4. Janfaza P, Nadol JB Jr. Temporal bone and ear. Janfaza P, Nadol JB Jr, Galla RJ, Fabian RL, Montgomery WW (eds.). Surgical Anatomy of the Head and Neck. Cambridge, Mass: Harvard University Press; 2011, pp.419-80.
5. Dieterich M, Brandt T. Vestibular system: anatomy and functional magnetic resonance imaging. Neuroimaging Clin N Am. 2001;11(2):263-73, ix.
6. Mei X, Atturo F, Wadin K, Larsson S, Agrawal S, Ladak HM, et al. Human inner ear blood supply revisited: the Uppsala collection of temporal bone-an international resource of education and collaboration. Ups J Med Sci. 2018;123(3):131–42. DOI:10.1080/03009734.2018.1492654. Epub 2018 Sep 11.
7. Mandalà M, Santoro GP, Awrey J, Nuti D. Vestibular neuritis: recurrence and incidence of secondary benign paroxysmal positional vertigo. Acta Otolaryngol. 2010;130(5):565-7. DOI: 10.3109/00016480903311278.
8. Gans RE: Evaluating the Dizzy Patient: Establishing Clinical Pathways. Hearing Review (1999); 6 (6): 45-47.
9. Wuyts FL, Van Rompaey V, Maes LK. "SO STONED": Common sense approach to the evaluation of the dizzy patient. Front Surg. 2016;3:32. DOI: 10.3389/fsurg.2016.00032.
10. Kattah JC, Talkad AV, Wang DZ, Hsieh YH, Newman-Toker DE. HINTS to diagnose stroke in the acute vestibular syndrome three-step bedside oculomotor examination more sensitive than early MRI diffusion-weighted imaging. Stroke. 2009;40(11):3504-10. DOI: 10.1161/STROKEAHA.109.551234. Epub 2009 Sep 17.
11. Fife TD, Satya-Murti S, Burkard RF, Carey JP. Vestibular evoked myogenic potential testing: Payment policy review for clinicians and payers. Neurol Clin Pract. 2018;8(2):129-34. DOI: 10.1212/CPJ.0000000000000430.
12. Korres S, Gkoritsa E, Giannakakou-Razelou D, Yiotakis I, Riga M, Nikolpoulos TP. Vestibular evoked myogenic potentials in patients with BPPV. Med Sci Monit. 2011;17(1):CR42-7.
13. Marzo SJ, Leonetti JP. The importance of magnetic resonance imaging in the evaluation of vertigo and imbalance. Skull Base Surg. 2000;10(4):171-2.
14. Walker MF. Treatment of vestibular neuritis. Curr Treat Options Neurol. 2009;11(1):41-5.
15. Marill KA, Walsh MJ, Nelson BK. Intravenous Lorazepam versus dimenhydrinate for treatment of vertigo in the emergency department: a randomized clinical trial. Ann Emerg Med. 2000;36(4):310-9.
16. Epley JM. The canalith repositioning procedure: for treatment of benign paroxysmal positional vertigo. Otolaryngol Head Neck Surg. 1992;107(3):399-404.
17. Brandt T, Steddin S, Daroff RB. Therapy for benign paroxysmal positioning vertigo, revisited. Neurology. 1994;44:796-800.
18. Sánchez Gil-Hernández Urtiz A, Herrera-Ortiz A, Perez-Cisneros MA. Study of the effectiveness of the liberatory manoeuvre in Lindsay-Hemenway syndrome. Rev Med Hosp Gen Méx. 2016;79(4):194-9. DOI: 10.1016/j.hgmx.2016.05.017.
19. Cetin YS, Omer OA, Demir UL, Kasapoglu F, Basut O, Coskun H. Comparison of the effectiveness of Brandt-Daroff Vestibular training and Epley Canalith repositioning maneuver in benign Paroxysmal positional vertigo long term result: A randomized prospective clinical trial. Pak J Med Sci. 2018;34(3):558-63. DOI: 10.12669/pjms.343.14786.
20. Goudakos JK, Markou KD, Psillas G, Vital V, Tsaligopoulos M. Corticosteroids and vestibular exercises in vestibular neuritis. Single-blind randomized clinical trial. JAMA Otolaryngol Head Neck Surg. 2014;140(5):434-40.
21. Fallahzadeh Abarghuei A, Fadavi-Ghaffar M, Tousi S, Amini M, Salehi AR. Effect of cawthorne and cooksey exercises on balance and quality of life of 60 to 80 year- old individuals in Shiraz: A randomized clinical trial. Med J Islam Repub Iran. 2018;32:74. DOI: 10.14196/mjiri.32.74.
22. Cohen HS. Disability and rehabilitation in the dizzy patient. Curr Opin Neurol. 2006;19(1):49–54.