Unilateral Vertical Nystagmus: The Heimann-Bielschowsky Phenomenon

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Objective: *To present clinical feature of Heimann-Bielschowsky phenomenon and strabismus association.*
Design: *Retrospective case series.*
Material and Method: *Medical records of cases between January 2004 and January 2008 were reviewed. Eight cases met the criteria. Age, sex, visual acuity, and eye examination data were collected.*
Results: *Six cases were male and two cases were female. Age at presentation ranged from 4 to 45 years old. Seven cases had vision of 6/60 or worse. Only one case had visual acuity 6/36. Strabismus was seen in five cases (exotropia 4 cases and esotropia 1 case). No one had symptoms of diplopia or oscillopsia. All cases had unilateral coarse, slow, pendular vertical oscillations occurring in the poor vision eye.*
Conclusion: *Monocular vertical oscillation may develop in an eye with reduced vision. It may be a common condition but under diagnosed.*

Keywords: *Eye movements, Heimann-Bielschowsky phenomenon, Nystagmus, Pathologic, Vertical oscillation, Low vision*

J Med Assoc Thai 2009; 92 (3): 373-6
Full text. e-Journal: *http://www.mat.or.th/journal*

The Heimann-Bielschowsky phenomenon (HBP) is a monocular vertical nystagmus. The oscillation is characterized by slow, coarse, pendular, variable amplitude movements in a profound visual loss eye. The mechanism of these movements is unknown but may be associated with disruption of the fusional vergence mechanism or the monocular visual stabilization system[1]. It may be an underdiagnosed condition in clinical practice.

The HBP had been first described in 1902 by Ernst Heimann. He stated that this nystagmus could occur in strabismic amblyopia and in some neurological diseases such as neurosyphilis, multiple sclerosis, epilepsy, and severe hysteria[2]. In 1931 Bielschowsky pointed out the difference between HBP and dissociated vertical deviation (DVD) movements and shown that amblyopia can cause unilateral nystagmus[3,4].

In 1979, Yee et al reported on 10 patients with profound visual loss and unilateral vertical nystagmus[5]. All their patients had a visual acuity (VA) of 6/60 or worse in the affected eye. The onset of their visual loss had ranged from birth to 40 years old. From the study, they found that the frequency of the vertical movement was less than 1 Hz, the amplitude usually less than 5° and the velocity less than 25° per second.

Smith et al had reported the Heimann-Bielschowsky Phenomenon eye movement frequencies of between 1 and 5 Hz, with amplitudes usually of 2° to 5° (but up to 30°)[6,7].

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Material and Method

Medical records of patients who visited between January 2004 and January 2008 were reviewed. Patients with monocular visual loss and unilateral vertical oscillation were included in the present study. Patients with bilateral vertical oscillation, unilateral horizontal oscillation, spasmus nutans, and incomplete medical records were excluded.

Demographic data, visual acuity, ophthalmologic pathology, ocular deviation, and characteristic of vertical oscillation were recorded.

Results

Eight patients met the criteria. Six cases were male and two cases were female. Age at presentation ranged from 4 to 45 years old. Seven cases had visual acuity of 6/60 or worse in the affected eye. Only one case had visual acuity of 6/36 in the affected eye. This case had anisometropic amblyopia. All patients had unilateral slow, coarse, variable amplitudes, pendular vertical movements in the poor vision eyes. Strabismus was seen in five cases (exotropia 4 cases and esotropia 1 case). No one had symptoms of diplopia or oscillopsia. Data of all patients are summarized in Table 1.

Discussion

The HBP may be a common but under diagnosed condition. It is important to keep this condition in mind in order to spare unnecessary neurological imaging and other investigations. It is necessary to observe this eye movement at least 1 minute to demonstrate the slow, pendular, vertical oscillation[8].

The present study showed eight cases of HBP. Three cases had poor vision from congenital causes such as anterior segment dysgenesia, microphthalmia, optic nerve hypoplasia, infantile cataract, and sclerocornea and five cases had acquired causes of poor vision (amblyopia, traumatic cataract, and absolute glaucoma). All cases had visual acuity of 6/36 or worse. Five cases had strabismus. Most cases with HBP had acuity of 6/60 or worse, but Davey et al[8] reported two patients with 6/9 (with contact lens corrected) acuity in the HBP eyes.

The mechanism of slow vertical oscillation in HBP is unknown. Pratt-Johnson and Tilson[9] postulated that HBP might be related to 'Central Fusion Disruption'. Leigh, et al[1] had proposed the pathogenesis of these eye movements that may relate

Table 1. Clinical data of the patients

Patients	Gender	Diagnosis/Lesions	Age at presentation (yrs)	Visual acuity		Ocular deviation	Characteristic of oscillation
				Right eye	Left eye		
1	M	Anterior segment dysgenesis, microphthalmos LE	4	6/9	Hm	Orthotropia	Coarse, slow, pendular, vertical movement
2	M	Penetrating injury, traumatic cataract RE, sensory XT, DVD	45	Hm	6/6	Exotropia	Coarse, slow, pendular, vertical movement
3	F	Optic nerve hypoplasia RE, sensory XT, IOOA	22	1/60	6/9	Exotropia	Coarse, slow, pendular, vertical movement
4	M	Strabismic amblyopia RE	20	Hm	6/6	Esotropia	Coarse, slow, pendular, vertical movement
5	M	Anisometropic amblyopia LE	45	6/6	6/36	Orthotropia	Coarse, slow, pendular, vertical movement
6	M	Sclerocornea, infantile cataract LE	4	6/9	Fc 1'	Orthotropia	Coarse, slow, pendular, vertical movement
7	F	Traumatic cataract RE	11	4/60	6/6	Exotropia	Coarse, slow, pendular, vertical movement
8	M	Absolute glaucoma LE	21	6/9	NPL	Exotropia	Coarse, slow, pendular, vertical movement

M = male, F = female, RE = right eye, LE = left eye, XT = exotropia, IOOA = inferior oblique overaction, DVD = dissociated vertical deviation, Hm = hand motion, Fc 1'= finger count 1 foot, NPL = no light perception

to disruption of steady gaze following monocular visual loss. They had attributed the HBP mechanism to first, may not relate to the 'common neural integrator' (gaze-holding network), second, 'fluctuations in the yoking mechanism (disconjugate saccades) and the third possibility, the abnormal vergence system. Another possible mechanism is a monocular stabilization system. Miles et al[10] had a study in rhesus monkeys and suggested that the ocular stabilization system depends on early connections in the visual pathways before inputs from the two eyes merging. Therefore, both abnormalities of vergence and monocular stabilization might contribute to the instability of gaze and cause vertical oscillation in monocular visual loss eye.

Some monocular visual loss patients have strabismus or manifested latent nystagmus in the affected eye, but some have HBP.

The differential diagnosis of unilateral nystagmus includes spasmus nutants and spasmus nutans-like disease[11], translated myokymia of the lower eyelid[12] and chiasmal tumor[13]. The eye movement of HBP is quite different to that seen in chiasmal tumors or in spasmus nutans. The eye movement of HBP is characterized by slow, coarse, pendular, variable amplitude movements but that in spasmus nutans is a small amplitude high-frequency type of oscillation.

The presented patients did not complain of oscillopsia but Rahman[14] reported one case with acquired symptomatic HBP who improved dramatically in oscillation after oral gabapentin treatment.

Heimann[2] and Smith[6] reported cases of improvement of oscillations following strabismus surgery but Davey[8] did not observe any definite reduction in oscillation in his patients after strabismus surgery.

Yee et al[5] mentioned one case with light perception acuity and senile cataract in affected eye that the HBP disappeared in this case after cataract surgery and correction with an aphakic contact lens. From vision-improving surgery such as cataract surgery, Davey[8] did not see any improvement in nystagmus and some cases developed vertical diplopia.

In conclusion, the HBP may be an underdiagnosed condition in an eye with decreased vision. It is associated with strabismus. The causes of visual loss in HBP may be congenital or acquired types. Symptoms of diplopia and oscillopsia are uncommon. Some cases with HBP improved of nystagmus and oscillopsia after medication treatment or strabismus and vision-improving surgery but some cases developed vertical diplopia.

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ภาวะลูกตาสั่นตาเดียวในแนวตั้ง (Heimann-Bielschowsky Phenomenon)

ธรรมนูญ สุรชาติกำธรกุล, พิทยา ภมรเวชวรรณ

วัตถุประสงค์: *เพื่อศึกษาลักษณะการเคลื่อนที่ของภาวะลูกตาสั่นตาเดียวในแนวตั้ง และภาวะตาเหล่ที่เกี่ยวข้อง*
วัสดุและวิธีการ: *เป็นการศึกษาย้อนหลัง จากเวชระเบียนผู้ป่วย ตั้งแต่เดือนมกราคม พ.ศ. 2547 ถึง เดือนมกราคม พ.ศ. 2551 พบผู้ป่วย 8 ราย ข้อมูลที่บันทึกมี อายุ เพศ ระดับสายตา การตรวจตา*
ผลการศึกษา: *พบผู้ป่วยชาย 6 ราย ผู้ป่วยหญิง 2 ราย อายุที่มาพบแพทย์ ตั้งแต่ 4 ปี - 45 ปี ผู้ป่วย 7 รายมีระดับสายตาน้อยกว่าหรือเท่ากับ 6/60 มี 1 ราย ที่มีระดับสายตา 6/36 พบภาวะตาเหล่ในผู้ป่วย 5 ราย โดย 4 ราย เป็นภาวะตาเหล่ออก และ 1 รายเป็นภาวะตาเหล่เข้า ไม่มีผู้ป่วยรายใดที่มีภาวะเห็นภาพซ้อนหรือภาพสั่น ผู้ป่วยทุกรายมีการขยับเคลื่อนที่ขึ้น-ลง ในแนวตั้งอย่างช้า ๆ ในตาข้างที่มัว*
สรุป: *ภาวะลูกตาสั่นตาเดียวในแนวตั้ง สามารถพบได้ในตาข้างที่มีตามัว ซึ่งอาจจะเป็นภาวะที่พบได้บ่อยครั้ง แต่ไม่ได้รับการนึกถึงเท่าที่ควร*