Master di II Livello in Vestibologia Pratica

Nistagmo/vertigine posizionale centrale

Mario Faralli

BPPV: Pattern tipico (Oculomotorio e Temporale)

CSP

- Latenza
- Parossismo
- Durata
- Dissociazione
- Geotropismo
- Inversione
- Esauribilità
- Ripetibilità
- Faticabilità

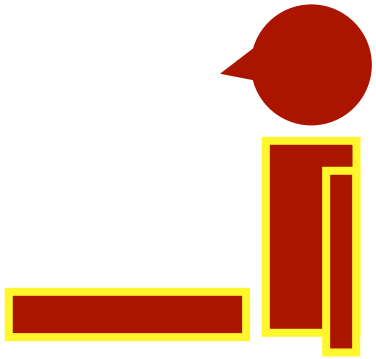
CSL

- Latenza (minore)
- Coniugato
- Non faticabile
- Durata > rispetto BPPV CSP
- Ny > intenso lato colpito (geo)
- Ny > intenso lato sano (apogeo)

CSA

- Genesi aspecifica
- latenza
- dissociazione
- Parossismo (poco)
- Esauribiltà
- Inversione (non sempre)
- faticabilità
- Sintomi (diversi)

DBN vs pDBN



DBN

Primary gaze
Sitting position

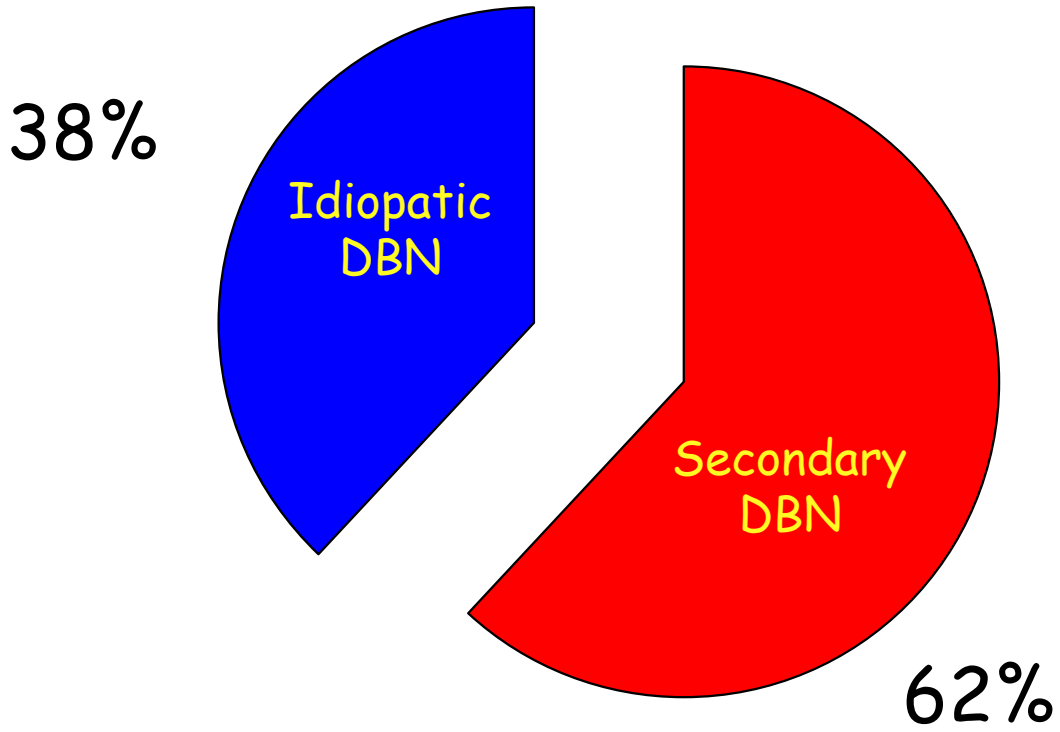
Supine position
Roll head, Dix-Hallpike
Head Hanging position

pDBN

DBN

- Slow phase velocity increases on lateral gaze (77%)
- Slow phase velocity increases on downward and decreased on upward gaze (idiopathic DBN: 64%; secondary DBN:82%)
- Convergence enhanced SPV (64%)
- SPV decreased/increased when patients was supine
- SPV increased in prone position
- Intermittent (upbeat)
- DBN is often associated with other oculomotor disorders (predominantly smooth pursuit deficits and impairment of the optokinetic reflex and visual fixation suppression of the vestibulo-ocular reflex (VOR)
- A frequent finding was unilateral or bilateral vestibulopathy
- Cerebellar signs (limb ataxia, dysarthria) were frequently seen (secondary DBN group)
- The most common presenting symptom was unsteadiness of gait (idiopathic 89%; secondary 81%) and oscillopsia (idiopathic 44%; secondary 38%)

Aetiology of DBN



Degenerative 20%
Vascular 9 %
Malformation 7%
Toxic 4%
Infiammatory 3%
Neoplastic 3%
Episodic ataxia 3%
Paraneoplastic 3%
Demyelinating 3%
Vestibular migraine 3%
B12 deficiency 2%
Traumatic 1%
Anti-GAD-associated 1%
Hypoxic 1%

Downbeat nystagmus: aetiology and commorbidity in 117 patients
J N Wagner, M Glaser, T Brandt, M Strupp
J Neurol Neurosurg Psychiatry 2008;79:672-677

pDBN is known to occur in CNS disease, particularly posterior fossa lesions

Nylen CO.
A clinical study on positional nystagmus in cases of brain tumor. Acta Otolaryngol 1931;(suppl 15):1–113.

Cawthorne T, Hinchcliffe R.
Positional nystagmus of the central type as evidence of subtentorial metastases.
Brain 1961;84:415–26.

Barber HO.
Positional nystagmus.
Otolaryngol Head Neck Surg 1984;92:649–55.

Kattah JC, Kolsky MP, Luessenhop AJ.
Positional vertigo and the cerebellar vermis.
Neurology 1984;34:527–9.

Positional down beating nystagmus in 50 patients: cerebellar disorders and possible anterior semicircular canalithiasis

P Bertholon, AM Bronstein, RA Davies, P Rudge, KV Thilo

J Neurol Neurosurg Psychiatry 2002; 72: 366-372

50 consecutive patients

Inclusion criteria

pDBN (not concomitant DBN)
Dix-Hallpike manoeuvre
head hanging position

torsional or horizontal nystagmic components added to pDBN were included

Exclusion criteria

Patients with DBN in any position of gaze while upright

CNS group:
38 patients

CNS dysfunction
38 patients; mean age 51, range 17 to 75 years old)

Idiopathic group:
12 patients

no evidence of CNS abnormality on clinical, oculographic, or imaging investigations (12 patients; mean age 50, range 33 to 78 years old

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P Bertholon, AM Bronstein, RA Davies, P Rudge, KV Thilo

J Neurol Neurosurg Psychiatry 2002; 72: 366-372

CNS group: 38 patients

Walking difficulties, falls
Orthostatic intolerance
Cerebellar abnormalties
Extrapyramidal disorders
Triggered by the Dix-Hallpike manoeuvre in 37 out of 38 patients;
bilaterally positive in 35 patients
Usually purely down beat (n=30)
no latency (n=37)
Short lived sometimes
No habituation

Idiopathic group: 12 patients

no evidence of CNS abnormality on clinical, oculographic, or imaging investigations.
History of migraine
History of VPPB
Trauma (one case)
Ear surgery (One case)
More positional symptoms
More torsional components
Latency (few seconds)
Habituation

Positional down beating nystagmus in 50 patients: cerebellar disorders and possible anterior semicircular canalithiasis

P Bertholon, AM Bronstein, RA Davies, P Rudge, KV Thilo

J Neurol Neurosurg Psychiatry 2002; 72: 366-372

CNS group: 38 patients

- Multiple system atrophy n° 13
- Cerebellar degeneration n°12
- Miscellaneous disorders n° 13

  Cerebrovascular disease, multiple sclerosis, Hydrocephalus.

Idiopathic group: 12 patients

- Migraine n° 5
- Middle ear surgery n° 2
- Myopathic syndrome n°1
- Recurrent posterior (BPPV) n° 2
- Positional vertigo after trauma n°1

Positional down beating nystagmus in 50 patients: cerebellar disorders and possible anterior semicircular canalithiasis

P Bertholon, AM Bronstein, RA Davies, P Rudge, KV Thilo

J Neurol Neurosurg Psychiatry 2002; 72: 366-372

CNS group: 38 patients

No patient had Arnold-Chiari malformation

Cerebellar flocculus is not the site
Responsible for transient pDBN

Nodular lesions in cats selectively cause similar transient pDBN

In patients with multiple system atrophy, the presence of pDBN was helpful in the differential diagnosis

Idiopathic group: 12 patients

The pDBN had characteristics wich suggested a peripheral labyrinthine disorder: vertigo, adaptation, and habituation

Spinocerebellar ataxia type 6 (SCA6)

Neurodegenerative disorder
Pure cerebellar ataxia syndrome
Late onset of symptoms
α_{1A} subunit of the P/Q type voltage-dependent calcium channel

Oculomotor abnormalities

- Gaze evoked nystagmus
- **Downbeat nystagmus**
- Saccade dysmetria

→ vertigo
→ oscillopsia

Neuronal loss is confined to Purkinje cells (granular cells, without any involvement of other central nervous system structures

Positional vertigo and macroscopic downbeat positioning nystagmus in spinocerebellar ataxia type 6 (SCA6)

Yabe I et al.
J Neurol (2003) 250: 440-443

Positional vertigo and macroscopic downbeat positioning nystagmus in spinocerebellar ataxia type 6 (SCA6)

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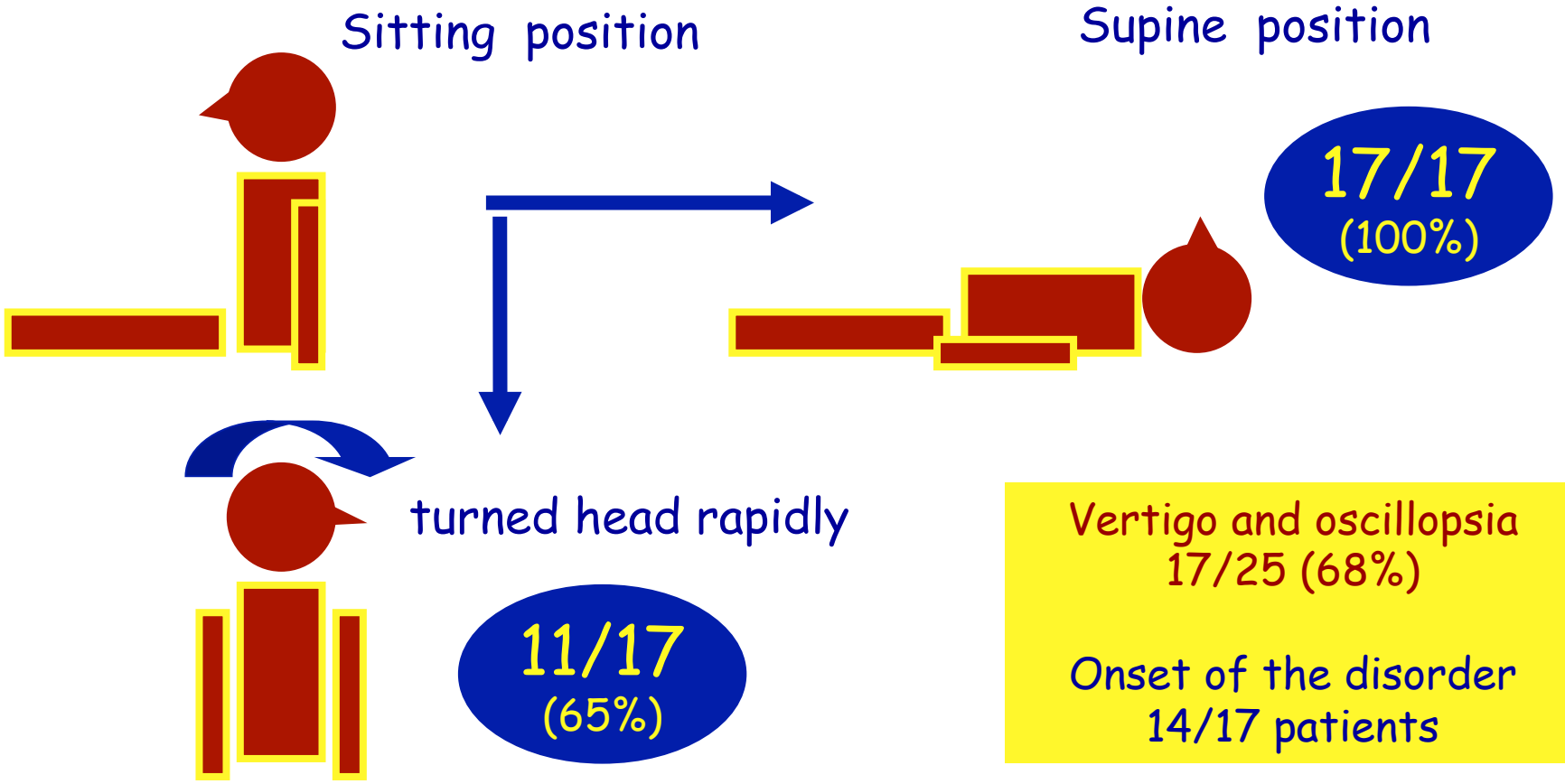
Clinical profiles of SCA patients



Vertigo and oscillopsia
17/25 (68%)

Onset of the disorder
14/17 patients

Positional vertigo and macroscopic downbeat positioning nystagmus in spinocerebellar ataxia type 6 (SCA6)

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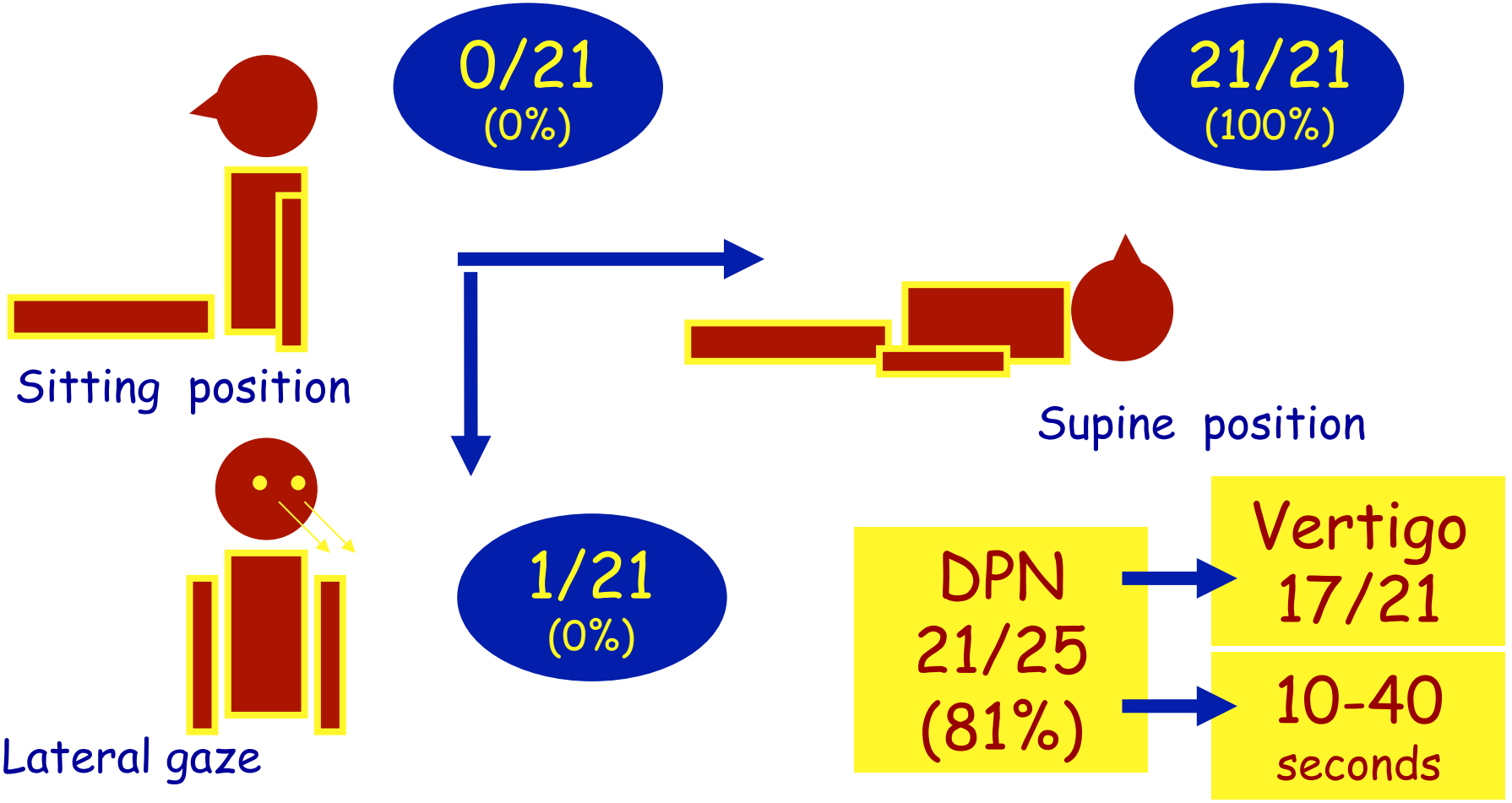
Clinical profiles of SCA patients



Positional vertigo and macroscopic downbeat positioning nystagmus in spinocerebellar ataxia type 6 (SCA6)

Yabe I et al.
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	SCA6 (n° 25)	SCA1 (n° 6)	SCA2 (n° 5)	SCA3/MJD (n° 16)	DCCA (n° 15)	MSA (n° 16)
Gender (man/women)	12/13	4/2	1/4	9/7	6/9	9/7
Age at onset	47.5 ±9	39.5 ±8	49.6 ±13	42.5 ±7.5	41.1 ±13	55.4 ± 8
Duration of disease	12.6 ±6	6.8 ±3.7	12.7 ±7	12.4 ±6.6	16.7 ±11	7.2 ±3
Score of ICARS	39.6 ±18	36 ±13	47 ±16	49.2 ±15.5	40 ±15	40 ±13
Gaze nystagmus	10 (44%)	0	0	15 (93.8%)	7 (46%)	0
Saccade dysmetria	22 (88%)	2 (33%)	1 (20%)	15 (93%)	12 (80%)	13 (81%)
Vertigo & oscillopsia	17 (68%)	0	0	4 (25%)	0	0
Downbeat positioning nystagmus	**21 (84%)**	**0**	**0**	**0**	**2 (13%)**	**1 (6.3%)**

DPN is a distinct clinical features in SCA6

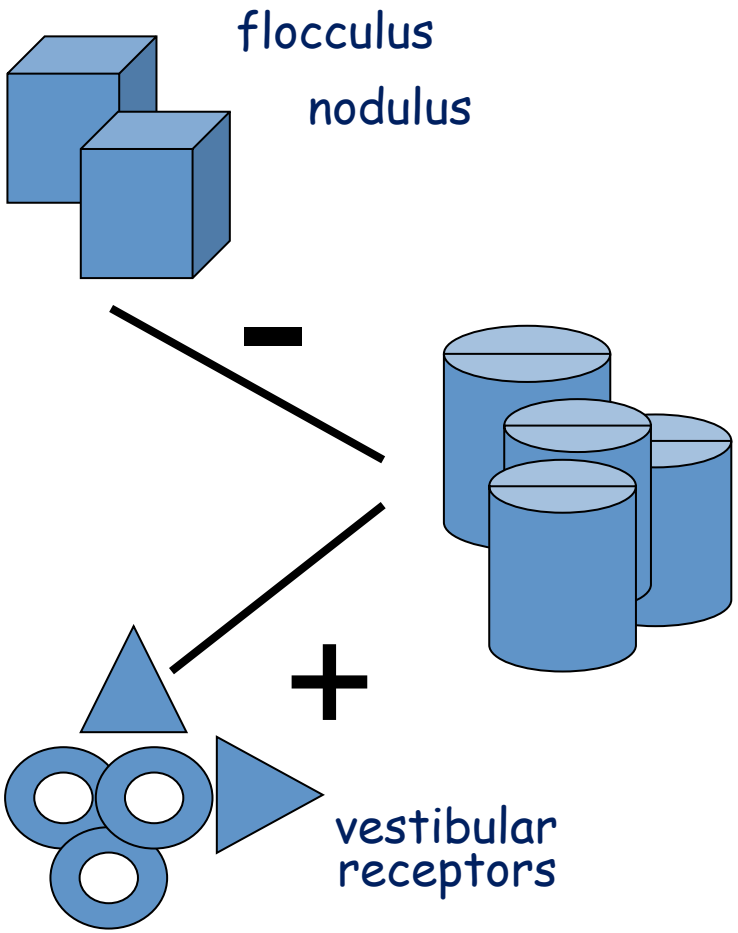
The symptoms were induced by DPN

Patients have DPN even at the onset

Purkinje cell loss is not uniform

Differences among each type of ataxia (SCA6 vs SCA3)

Role of Purkinje cells?

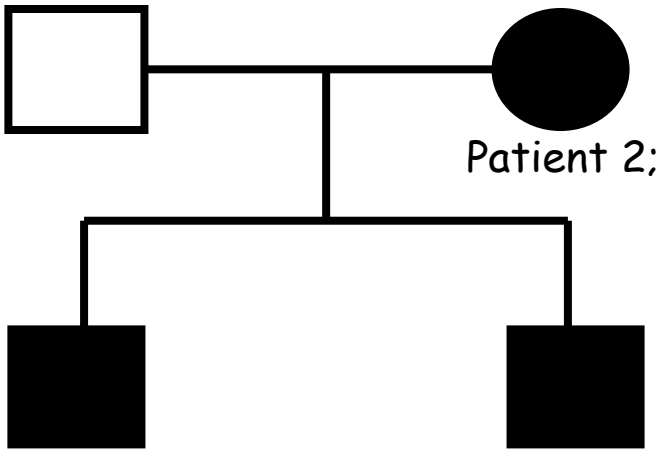


VOR

Stabilizes retinal images during head movement

Leigh RJ, Zee DS (1999) Nystagmus caused by central vestibular imbalance. In: Leigh RJ, Zee DS (eds) The Neurology of Eye Movements.Oxford Univ Press,New York, pp 415-424

Zee DS,Yamazaki A, Butler PH,Gucer G (1981) Effects of ablation of flocculus and paraflocculus of eye movements in primate. J Neurophysiol 46:878-899



Downbeat positioning nystagmus is a common clinical feature despite variable phenotypes in an FHM1 family *(Ichiro Yabe et al.)*

J Neurol (2008) 255: 1541-1544

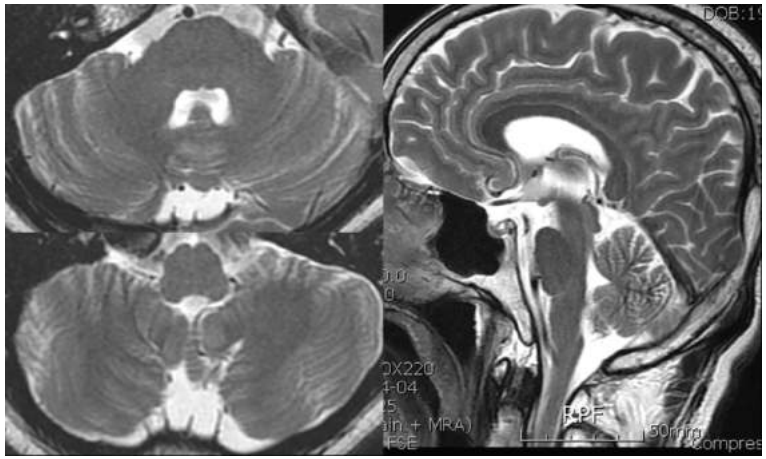
	Patient 1	Patient 2	Patient 3
Age at onset of main clinical manifestation	36	63	20
Age at onset of vertigo and oscillopsia	20	girlhood	teen's
Gaze nystagmus	moderate	mild	mild
Downbeat positioning nystagmus	severe	severe	severe
Cerebellar ataxia	moderate	mild	Very slight
Headache attack	+	-	+
Cerebellar atrophy	**moderate**	**moderate**	**moderate**
Main clinical manifestation	Hemiplegic migraine	Episodic ataxia	Migraine without aura

Downbeat, positional, and perverted head-shaking nystagmus associated with lamotrigine toxicity (Sun-Young Oh et al) J Clin Neurol 2006

Transient dysfunction of the vestibulocerebellum induced by drugs

Oculomotor abnormalities → 600mg/day

pDBN
pHSN
Lateral gaze

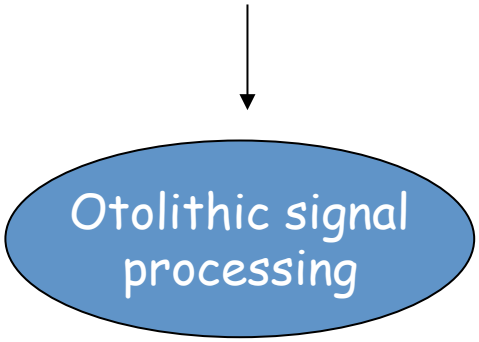


- Severe oscillopsia
- Dizziness
- Ataxia
- Vomiting, abdominal pain

2 cases: MRI

Positional nystagmus and vertigo due to a solitary brachium conjunctivum plaque
E Anagnostou, D Mandellos, G Limbitaki
J Neurol Neurosurg Psychiatry 2006; 77:790-792

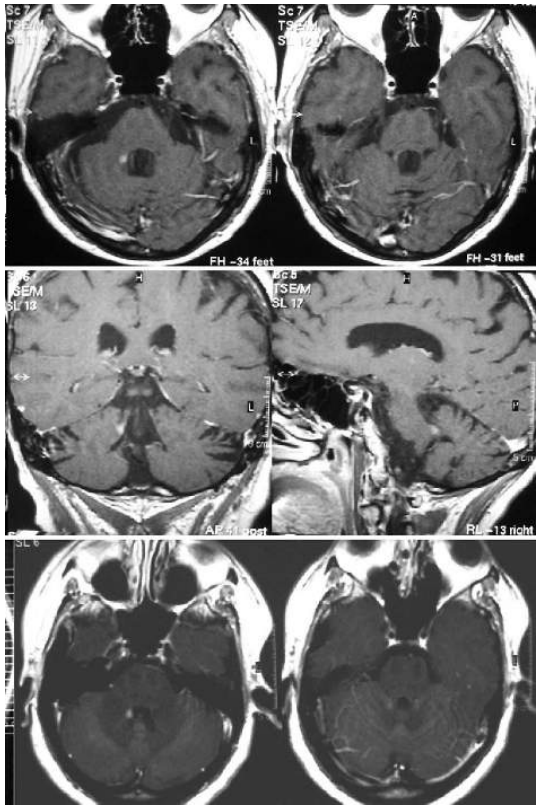
Solitary enhancing lesion located in the right brachium conjunctivum



Dynamic Otolithic syndrome

intense vertigo and persistent DBN upon tilting heads relative to gravity.

Symptoms stopped by bringing head back to the Upright position



pDBN and migraine

Episodic vertigo related to migraine (90 cases: vestibular migraine?)
Dieterich M, Brandt T.
J Neurol. 1999; 246: 883-892.

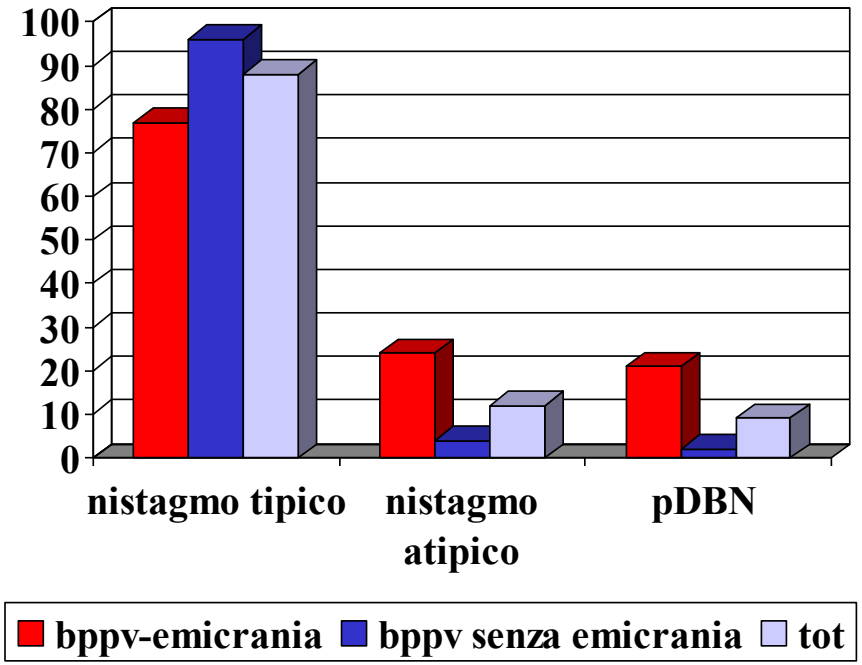
- Spontaneous vertigo (nystagmus)
- Spontaneous vertigo (ny) and positional vertigo (ny)
- **Positional vertigo (nystagmus)**

Von Brevern M, Radtke A, Clarke AH, Lempert T
Migrainous vertigo presenting as episodic positional vertigo.
Neurology 2004; 62:469-472

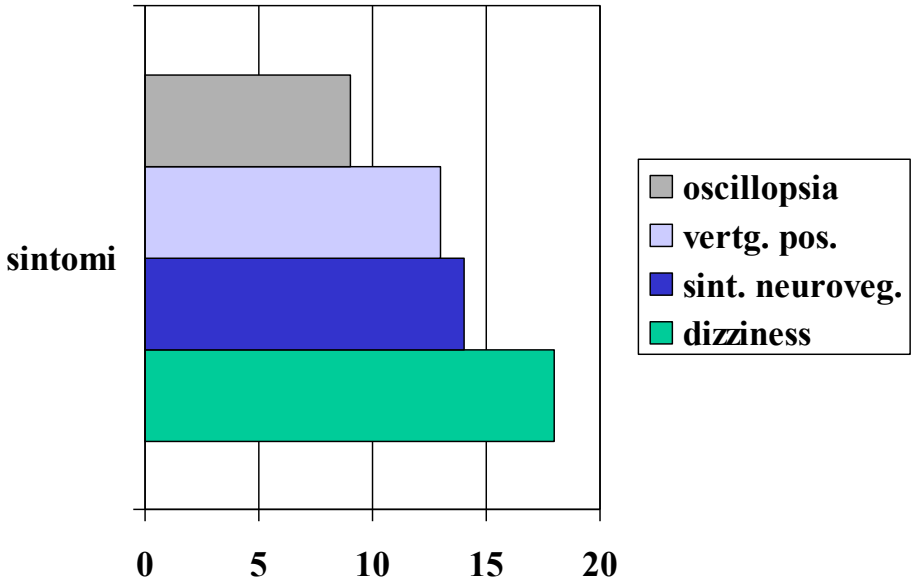
Ocular findings

- Spontaneous horizontal ny
- Spontaneous downbeating ny
- Spontaneous upbeating ny
- Pure torsional ny (?)
- **positional DBNy**
- positional horizontal ny (apogeo –geo) (!!)

pDBN and migraine
(esperienza personale)



Pazienti bppv	Ny atipico	pDBN
con emicrania	18/77 (23.3 %)	16/77 (20.7 %)
senza emicrania	4 /109 (3.6 %)	2/109 (1.8 %)
totali pazienti	22/186 (11,8 %)	18/186 (9.6 %)



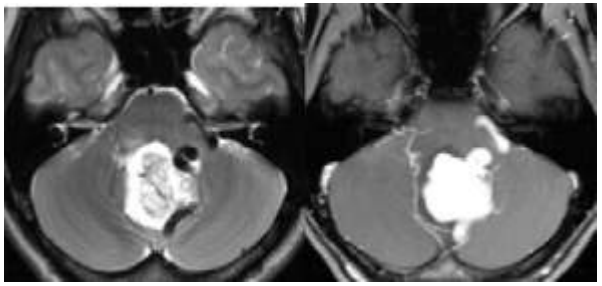
Ny assente in sitting position
Ny presente in posizione declive
Componente torsionale assente
Ny posizionale (non Ny posizionamento)
Periodo intercritico BPPV

Central positional nystagmus associated with cerebellar tumors: Clinical and topographical analysis.

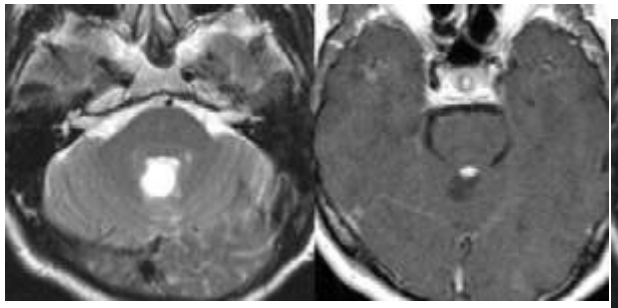
Cho BH[1], Kim SH[1], Kim SS[1], Choi YJ[2], Lee SH[3].

J Neurol Sci. 2017 Feb 15;373:147-151.

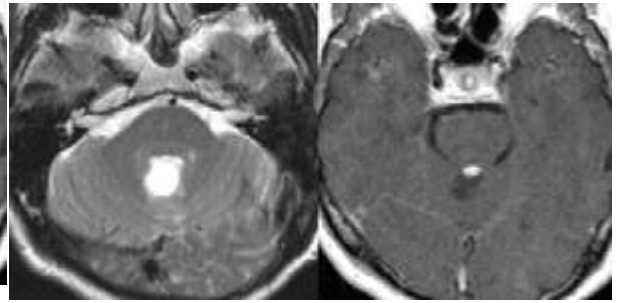
- 4 Patients
- Apogeotropic positional nystagmus during supine roll tests
- Initial diagnosis of BPPV (treated without success)
- Neurological examination normal (except positional ny)



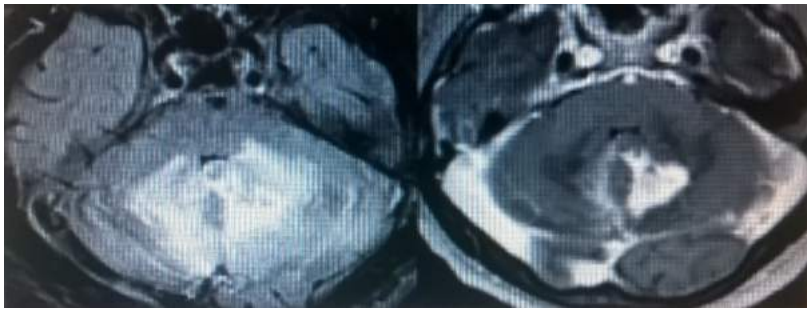
Hemangioblastoma



Hemangioblastoma



Metastatic breast cancer



Metastatic renal cancer

An apogeotropic Positional nystagmus may be an isolated finding in patients with a cerebellar tumor. especially when refractory to repeated canalith repositioning maneuvers.

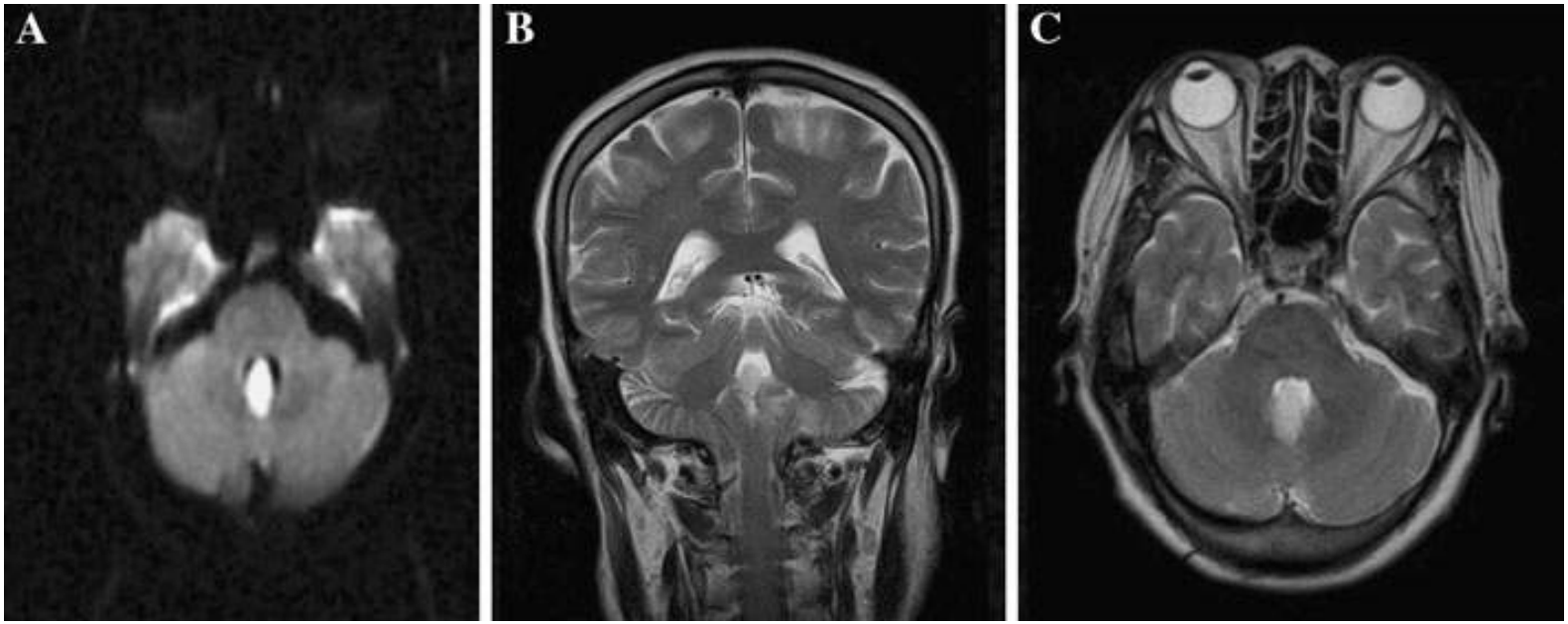
Apogeotropic central positional nystagmus as a sole sign of nodular infarction.

Kim HA[1], Yi HA, Lee H.

Neurol Sci. 2012 Oct;33(5):1189-91

Usually associated with

- Spontaneous or gaze-evoked nystagmus
- Perverted head-shaking nystagmus, cerebellar
- dysmetria
- severe gait ataxia with falling.



Geotropic central paroxysmal positional nystagmus in a patient with human immunodeficiency virus encephalopathy.

Yang TH[1], Oh SY.

J Neuroophthalmol. 2014 Jun;34(2):

- no spontaneous nystagmus
- gaze-evoked nystagmus (GEN)
- geotropic horizontal nystagmus (Positional tests)
- mild dysmetria
- intention tremor bilaterally
- severe ataxia.

Persistent geotropic positional nystagmus in unilateral cerebellar lesions

Seo Young Choi, Ji-Yeong Jang, Eun Hye Oh, Jae-Hwan Choi, Ji Yun Park, Seong-Han Lee, Kwang-Dong Choi

Neurology. September 11, 2018; 91 (11) Article

58 patients with persistent geotropic positional nystagmus

Seven patients with unilateral cerebellar lesions

The prevalence of central lesions in persistent geotropic positional nystagmus was 12% (7/58).

All patients showed impaired horizontal smooth pursuit bilaterally, and 3 of them also had positional downbeat nystagmus.

Conclusioni

- Un nistagmo (vertigine) posizionale può indicare una lesione SNC
- Valutare elementi di sospetto!
- Meccanismi patogenetici (pDBN e Apogeotropo)
- SM, Ischemie, intossicazioni, malformazioni, neoplastica, degenerazioni cervelletto (unica manifestazione)
- Lesioni dorsolaterali al 4° ventricolo, peduncoli cerebellari inferiori, verme dorsale, cerebellari diffuse