Vestibular Neurology. An Update

Jorge C Kattah, M.D.

University Of Illinois College of Medicine. Peoria.

Illinois Neurological Institute

OSF HealthCare

UIC UNIVERSITY OF ILLINOIS COLLEGE OF MEDICINE PEORIA

Background

- Currently the evaluation of patients with acute vestibular (AVS) or acute ataxia syndrome (AAS) varies with the facility specific diagnostic resources. *In-hospital and ED Video-Oculography (VOG) evaluation is available only in a few facilities, neuroimaging* in others. Few previous studies reported the specific contributions of the two diagnostic methods.
- Each test modality have limitations at the present time mostly because the membranous labyrinth cannot easily be imaged and the majority of *AVS* patients have a *peripheral etiology*. In addition, central structural imaging abnormalities may be *sub-radiological* with false negative imaging.
- *Clinical/VOG abnormalities* differentiate central from peripheral localization and do require *imaging for precise AVS/AAS etiology.* Here we explore VOG/clinical exam to guide imaging selection

Methods

- Beginning in January 2017 and ending in December 2021, we performed clinical, and VOG on AVS/AAS patients.
- We performed clinical exam and fixation block with the (***CHARTR 200 goggles***). If *spontaneous nystagmus* was present we did quantitative bedside recording of spontaneous nystagmus, saccade, pursuit and quantitative video head impulse test (vHIT) using ***Otometrics (NATUS Video-Impulse goggles***)
- If *spontaneous nystagmus was absent,* we performed positional maneuvers and treated the patient in accordance to the affected canal (canalolithiasis of posterior or horizontal canal) without further tests

TARGET EXAMINATION (Performed by neurologist/ENT/ audiologist/PT and vestibular technologist)

- Ideally examine the patient sitting up
- Observe the ability to remain seated without assistance
- Examine visual fixation: ***The patient fixates on a distant target***
- ***Examine the effect of fixation block (any method)***
- Examine **saccade** (both the ability to *generate the eye movement* and to *hold eccentric gaze*) pursuit, and vergence eye movements,
- Examine the dangerous D's: Diplopia, deafness, dysarthria , dysphonia, dysphagia, dysmetria
- Perform ***Head Impulse and test for skew***
- Look for obvious lateralizing signs (Weakness /limb ataxia/loss of sensation)
- Test posture and gait
- After this is complete, if normal, may proceed to *positional testing*

Bedside Video- Recording of the HIT and eye movements

Otometrics Natus Goggles

Interacoustic Goggles

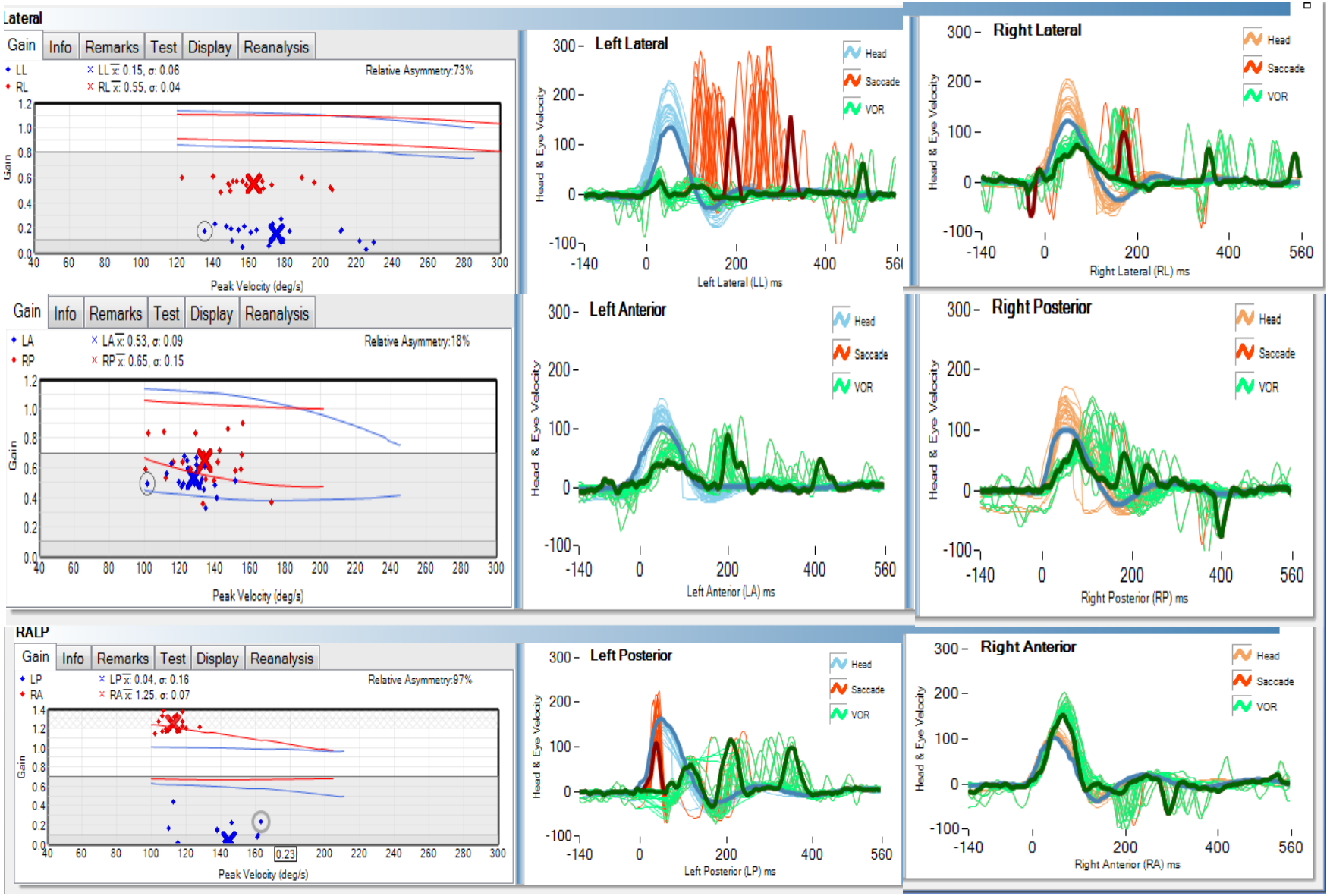
Video Goggles to record eye movement

QUANTITATIVE vHIT ANALYSIS

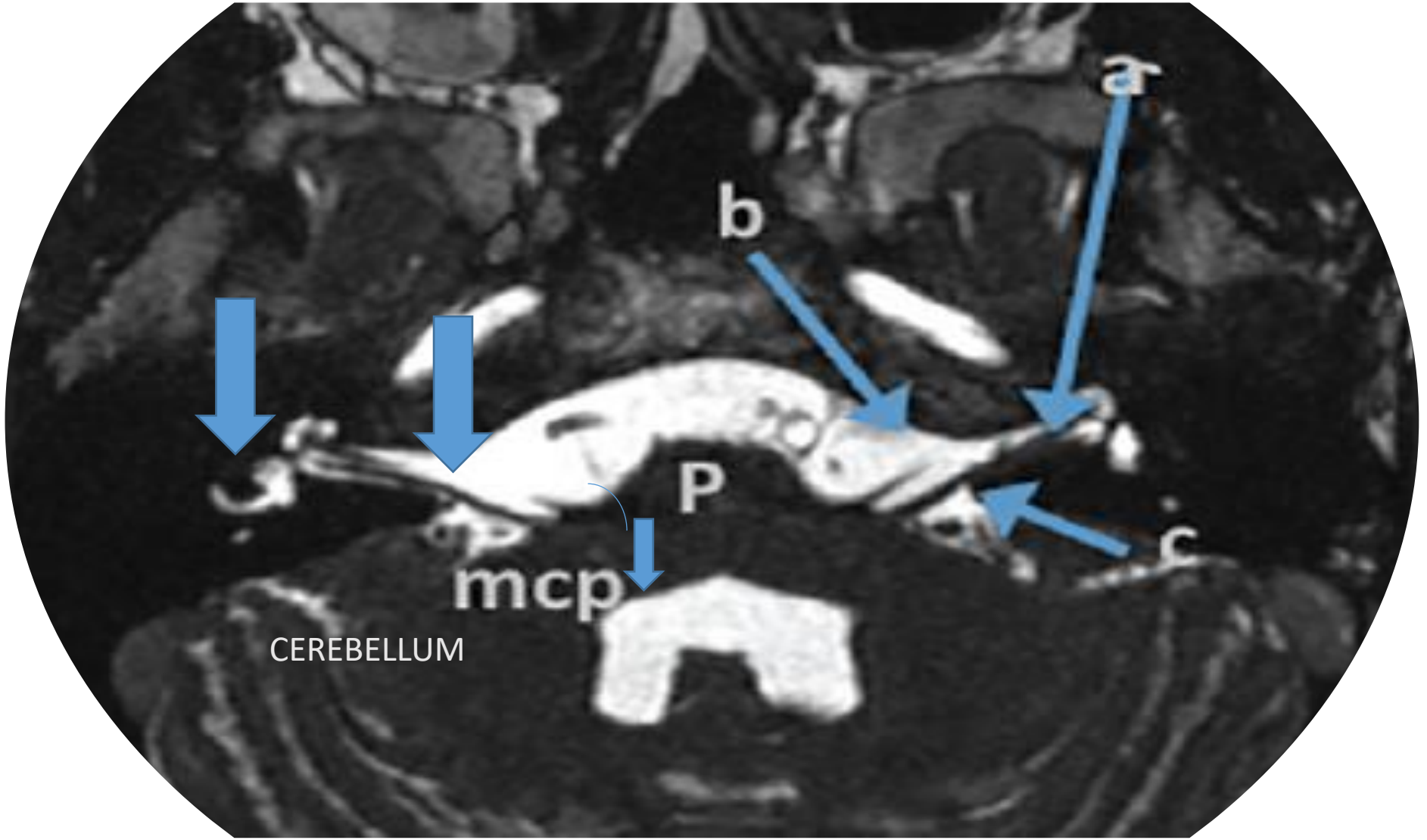
LL 0.15 RL 0.55

LA 0.53, RP 0.65

LP 0.16 RA 1.25



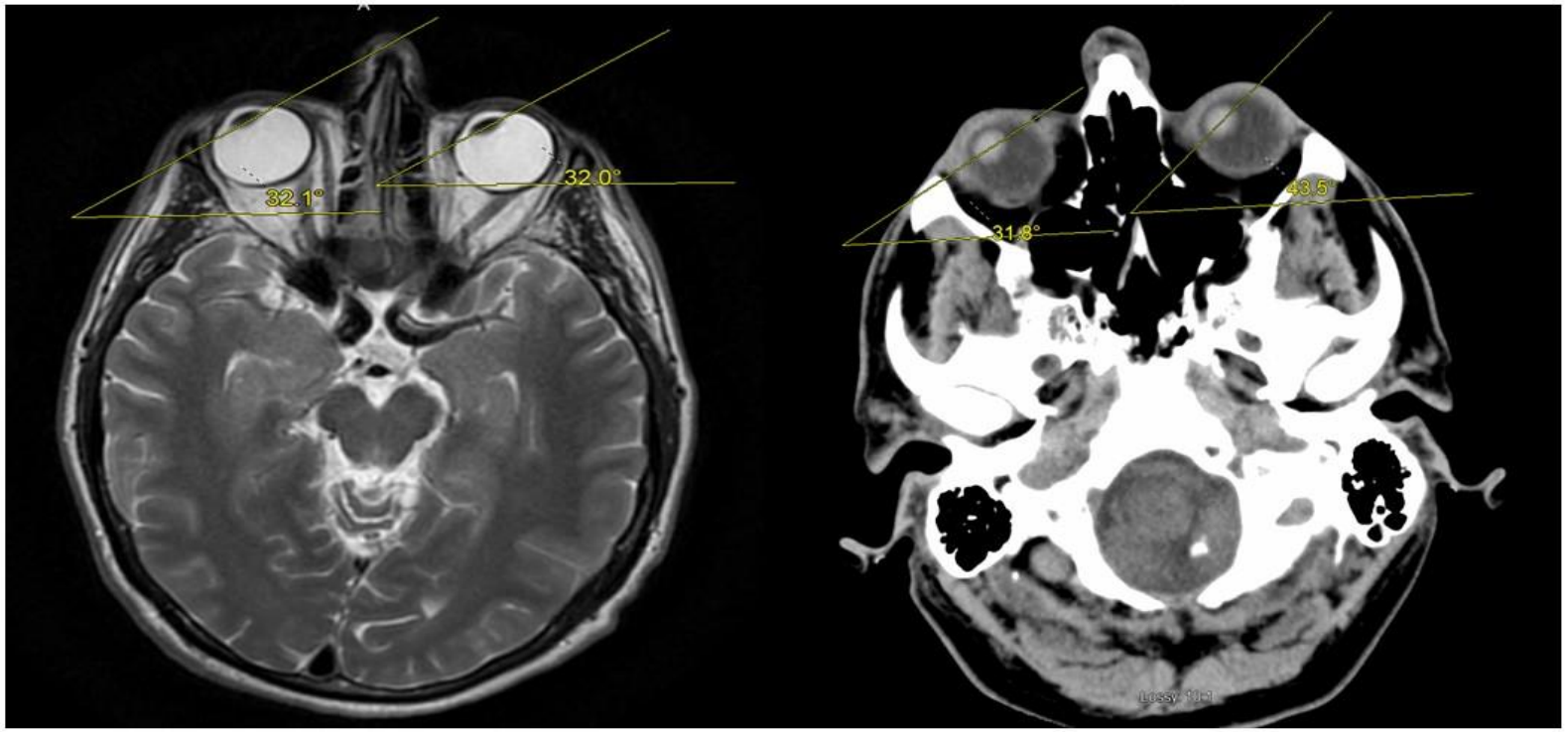
POTENTIAL LESION LOCALIZATION IN AVS



Neuroimaging. Retrospective Review (Not guided with input from exam or VOG results).

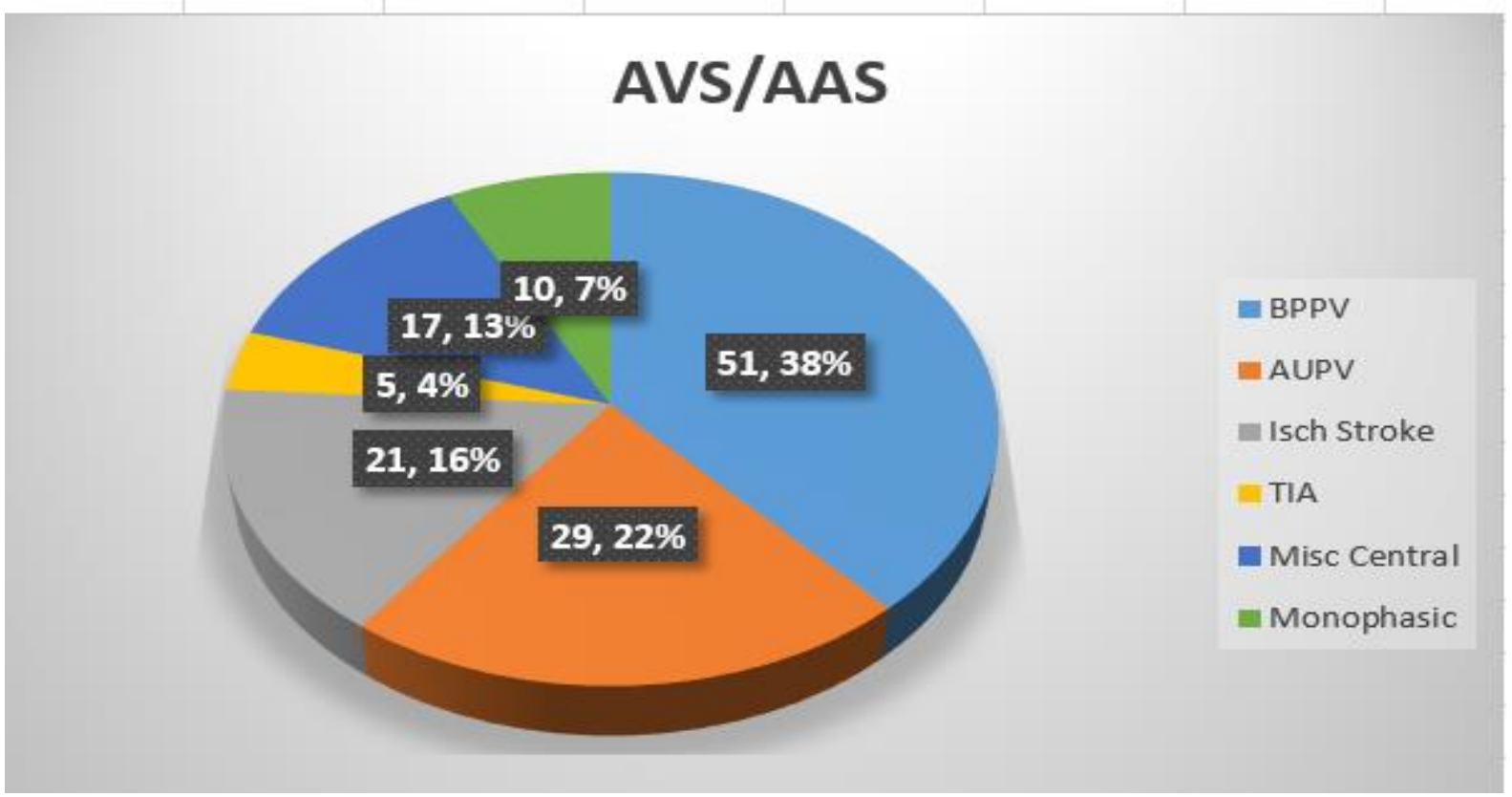
- All patients had MRI studies; in few, CT scan was an alternative for those patients who could not undergo MRI testing. The vestibular consultation took place after imaging in most (> 90%).
- MRI performed at our Institution involved the MRI unit that was first available, following a uniform MRI stroke protocol. We searched specifically for parenchymal imaging abnormalities diagnostic of ischemic stroke, hemorrhage and other central abnormalities.
- In addition, we looked for *horizontal conjugate gaze deviation (hCGD)*, preferably in T2 MRI sequences, we also looked in the CT scan, and in both studies if performed, we measured the degree of h- CGD if present, and compared it with normal controls
- We looked at the signal void of the vertebral arteries in cross- section T2 MRI sequences which we compared with MRA results.
- We then compiled the bedside and VOG evaluation with imaging

Radiographic Ocular Gaze Deviation (Rad- OLD) In the Acute Vestibular Syndrome (AVS)



Rad-OLD matches the slow phase of nystagmus and does not differentiate central versus peripheral vestibulopathy. ***Kattah J, Newman-Toker DE. Ann NY Acad Sci.*** 1233:(1) 249-255, **2011**

RESULTS: 133 AVS/AAS Patients



BPPV ABCD2 SCORE 3.70

- Benign paroxysmal positional vertigo (BPPV) was the most frequent (n=51): *posterior canal 33, horizontal canalolithiasis 14, horizontal cupulolithiasis 2*, and among the posterior canal, n= 1 was polycanalicular unilateral (horizontal/posterior), and n= 1 was bilateral simultaneous posterior canal
- Imaging in BPPV: not performed in n=6/51 patients, *n=18/51 had normal MRI of the brain*, *n=**18**/51 had non-specific periventricular leukoareosis, one with a left hemispheric stroke*). *N=1*/51 had multiple sclerosis (MS).
- ***N= 3/51 suffered recent TBI (post-traumatic BPPV)***: n=1 focal encephalomalacia, n=1 periorbital and facial edema and n=1 had post traumatic parenchymal and subdural hematoma; n= 4/51 had recent neurosurgery or other surgical procedures (one with a small, incidental tentorial meningioma), Finally, n=1 had amyloid angiopathy.

HORIZONTAL CANAL BPPV CHARACTERISTICS

- h -NYSTAGMUS BEATING IN A GEOTROPIC DIRECTION IN RESPONSE TO HEAD ROLL
- HIGH SLOW PHASE VELOCITY WHEN COMPARED WITH UNILATERAL VESTIBULOPATHY
- NOT ASSOCIATED WITH RADIOGRAPHIC HORIZONTAL NYSTAGMUS
- RESPONDED WELL TO CANAL REPOSITIONING

Unilateral Acute Vestibular Neuropathy (AUPV), previously Vestibular Neuritis ABCD 2 SCORE 3.70

- N= *28/29* acute unilateral vestibulopathy (AUPV) group underwent MRI, one had a CT scan only because of a non-compatible pacemaker. We found horizontal(h) conjugate deviation (h-CGD) of the eyes in the same direction to the slow phase of the h-nystagmus in *(82.7%).*
- It was the most common AUPV imaging finding (n=25/29), these patients had ipsilesional decreased h-VOR gain with catch up saccades in the VOG (and spontaneous horizontal/torsional, unidirectional nystagmus, n=4 did not have h-CGD. In contrast, *h-CGD* was present only in n=4/20 strokes (20%). *p value < .0001.*
- *Three patients with VZV* (Ramsey-Hunt) had an enhancing vestibular nerve, and one had a suppurative middle/inner ear fluid. *N=16 patients had leukoareosis.* We included in this group one patient with *vestibular paroxysmia* who had neurovascular compression

REASON/S TO COME UP WITH A SENSITIVE COMBINATION OF CLINICAL FINDINGS IN AVS PATIENTS

- THE ***HEAD IMPULSE TEST*** IS VERY HELPFUL IN LOCALIZING LESIONS RESPONSIBLE FOR *AN ACUTE VESTIBULAR SYNDROME*, IMPORTANTLY WHEN NYSTAGMUS IS PRESENT.
- *IF THE HORIZONTAL HIT IS NORMAL*, IT INDICATES THAT THE a VOR IS NORMAL, THUS PROVIDING UNEQUIVOCAL EVIDENCE THAT THE *LESION SPARES THE VESTIBULAR PERIPHERY*.
- THEREFORE, *A NORMAL HIT IN AVS SUGGESTS A PICA VASCULAR TERRITORY LOCALIZATION*, HOWEVER, SOME LESIONS INVOLVING THE AICA VASCULAR TERRITORY CAUSES A POSITIVE h-HIT.
- **THERE WAS THEN NEED FOR A SOLUTION TO THIS EVENTUALITY**

Stroke
JOURNAL OF THE AMERICAN HEART ASSOCIATION

American Stroke Association℠
A Division of American Heart Association

- Jorge C. Kattah, Arun V. Talkad, David Z. Wang, Yu-Hsiang Hsieh and David E.Newman-Toker
- **Bedside Oculomotor Examination More Sensitive Than Early MRI. HINTS to Diagnose Stroke in the Acute Vestibular Syndrome : Three-Step Prospective Study Of acute Vestibular Syndrome**
- ISSN: 1524-4628
- Print ISSN: 0039-2499. Online
- Stroke is published by the American Heart Association. 7272 Greenville Avenue, Dallas, TX 72514

doi: 10.1161/STROKEAHA.109.551234

Stroke 2009, 40:3504-3510: originally published online ***September 17, 2009***

Illinois Neurological Institute
OSF Saint Francis Medical Center

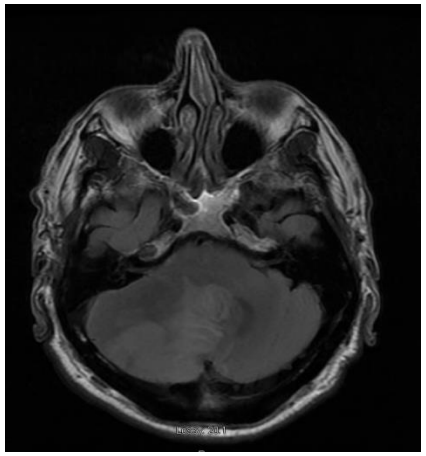
CENTRAL CAUSES: Ischemic Stroke ABCD2 Score : 3.70. TIA ABCD2 SCORE 4.00

- *Central causes included 20 ischemic vascular vertigo/ataxia patients*, with an *AVS*: n=12, *AAS* n=7, two acute cochlear-vestibular syndrome patients (possibly involving the labyrinth), n=1 small lacunae affecting the vestibular nucleus. N= 9 additional patients fit the Bárány Society criteria for the diagnosis of vestibular pathway TIA.
- N=3/20 (15%) had an initial *false-negative MRI.* Most of the strokes (*n=12*) ranged in size from 2 to 15 mm in their greatest diameter (lacunar). *Eight patients had large cerebellar, cerebello -medullary or bilateral cerebellar strokes*. One patient had a presumed labyrinth stroke along with an acute occipital lobe stroke. The cerebellar strokes were not complicated by ischemic edema. One patient had a basilar artery occlusion (BAO) with unilateral hearing loss; he remained stable without need for thrombolysis

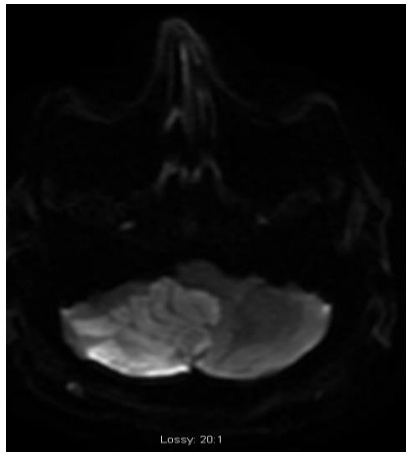
Direction Changing Nystagmus

09-19-2013

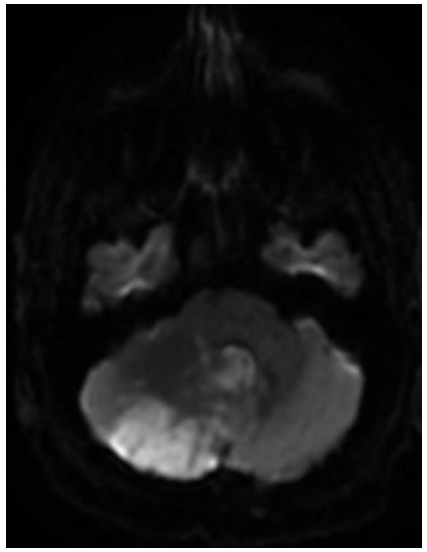
09-20-2013



09-23-2013



09-23-2013



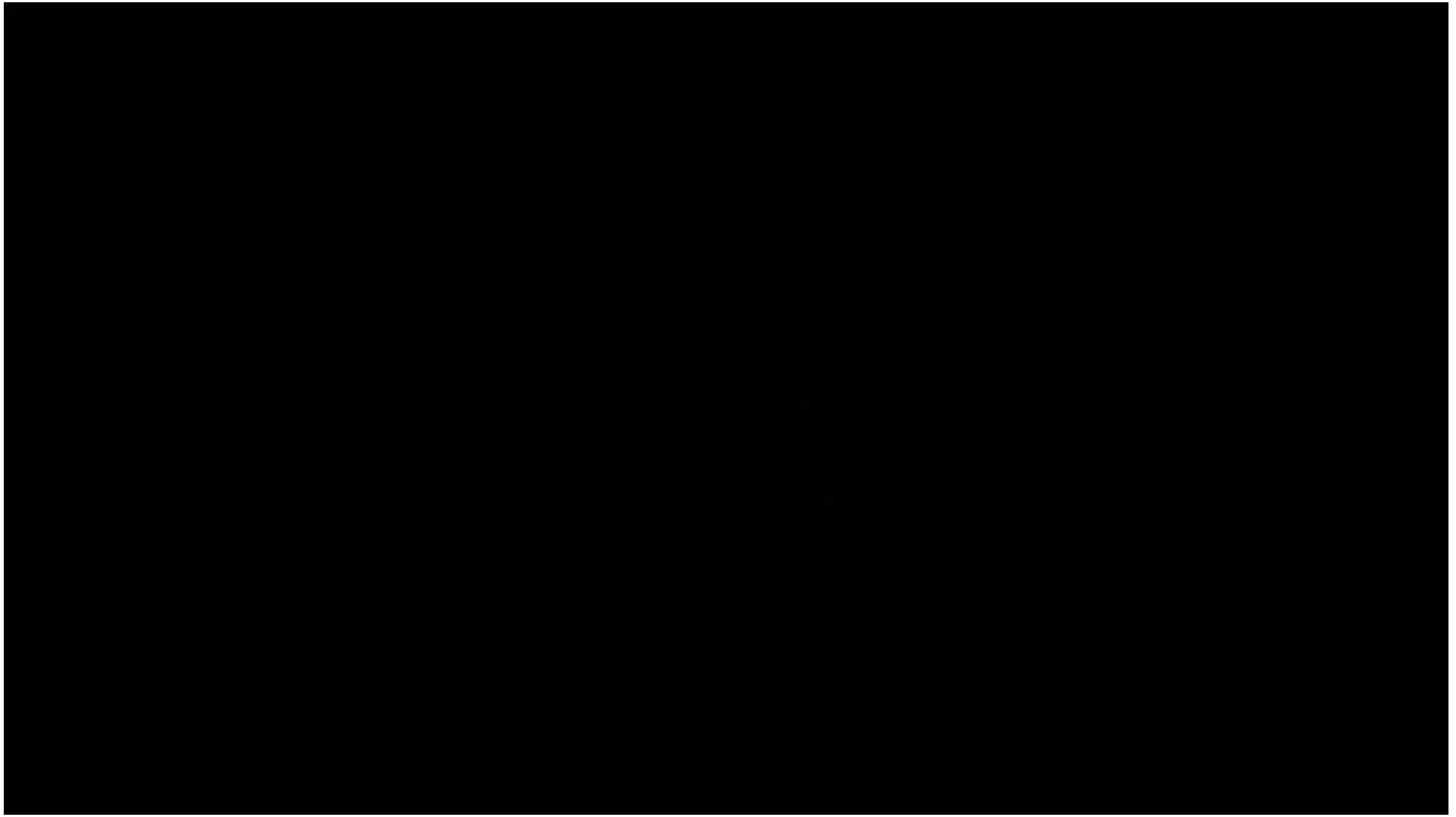
09-23-2013

09-25-2013

09-26-2013

09-30-2013

LARGE CEREBELLAR STROKE PRESENTING AS PSEUDONEURITIS



SKEW DEVIATION

DEFINITION: Vertical Misalignment of the eyes in te primary posiiton of gaze due to impairment of the otolith ocular reflfex (OTR).

MOST COMMON CAUSES OF SKEW DEVIATION:

- Brainstem Lesions : Often large amplitude, > than 3-prism diopters
- Peripheral vestibular Lesions: Often low amplitude, they do not cause diplopia .
- Cerebellar Lesions can cause it, and it may be alternating skew

AT TIMES IT MAY ASSOCIATED WITH HEAD TILT AND CONJUGATE OCUALR TORSION MEASURED WITH FUNDUS PHOTOGRAPHY. OCULAR TILT REACTION

Clinical/VOG: Nystagmus Findings Ischemic Stroke

- Primary gaze ***UBN:*** n=**4** (small brainstem strokes involving the dorsal medulla and pontomedullary junction), often mixed with different types of h- nystagmus, (gaze paretic in n=3).
- Three cerebello medullary or bilateral cerebellar strokes showed mixed horizontal/***DBN.***
- **Unidirectional horizontal (h)** *nystagmus i*n ***three*** lacunar stroke patients **(Two with normal h-VOR gain, one bilaterally decreased).**
- One patient had *apogeotropic positional h-nystagmus*
- One acute unilateral deafness + positional posterior canal BPPV, along with an acute occipital lobe infarct (presumably labyrinth infarct). One had positional UBN.
- Two patients had bilateral gaze paretic nystagmus.
- One medullary stroke had ocular laterodeviation (OLD).

VOG Findings. Ischemic Stroke

- Normal vHIT gain (central HIT) n= **11/21** + n= **3/21** clinically normal could not complete vHIT
- ***Bilateral low gain n=5/21! (25%)***
- ***Peripheral unilaterally low*** **n=2/21 (AICA strokes).** These two patients had acute unilateral deafness, and one had h-direction changing nystagmus.

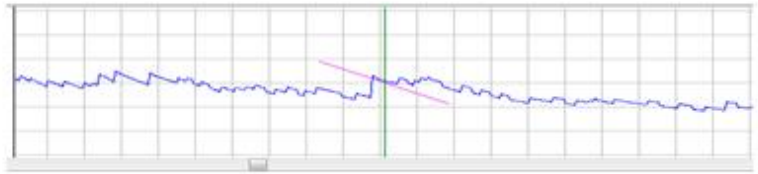
TIA n= 5 Fitting the Bárány Society definition for Intermittent Vascular Vertigo. ABCD2 Score 4.40.

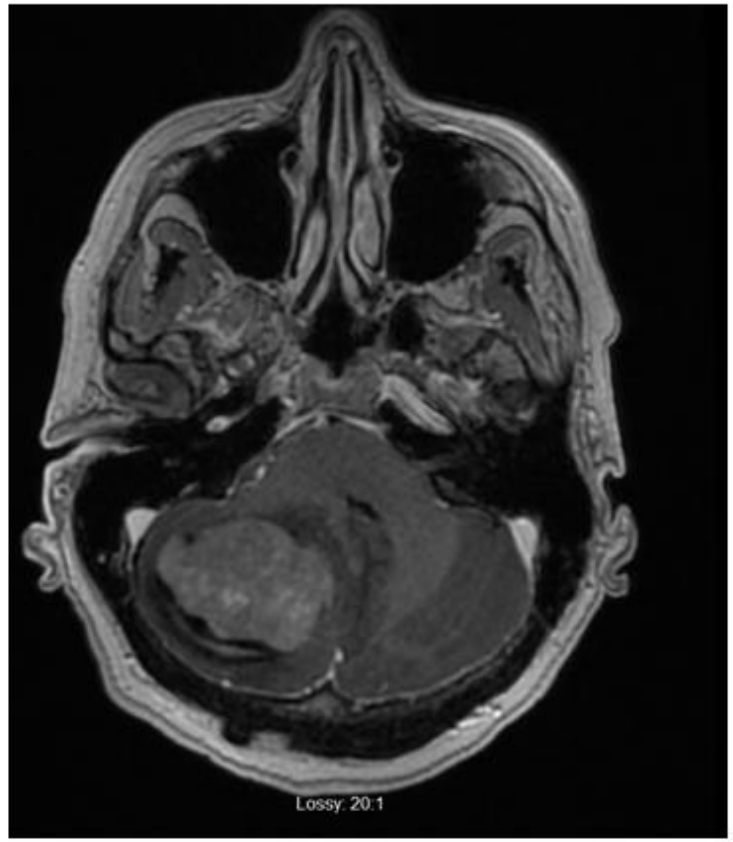
- The h- VOR gain was unilaterally decreased in n=1, bilaterally decreased in n=1 and normal in n=3 , with one showing a unilateral increased gain.
- Nystagmus was h-unidirectional in two patients with positional UBN, and UBN/DBN transition. One patient had fixation UBN, one had acute deafness and positional UBN, and one patient did not have nystagmus.
- None of the five TIA patients had imaging evidence of *acute stroke*. One had a ***left vertebral artery (VA) occlusion***, one had ***bilateral VA stenosis***, ***two*** had significant ***asymmetric, though patent VA*** , and one had ***VA fenestration***.

N=17 Miscellaneous Central Causes of AVS/ASS, n=7/17 had a normal MRI. ABCD2 Score: 2.35

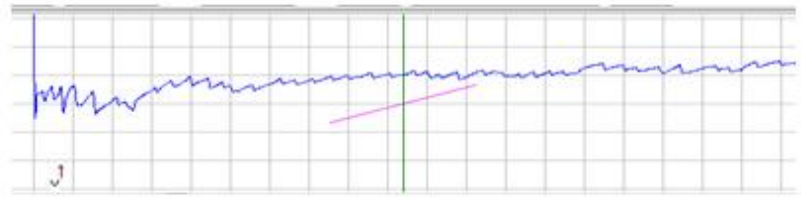
- Mass Lesions: n=5/17 (***3 brain tumors*** : Cerebellar Medulloblastoma, Bilateral Metastatic Lymphoma to pons, Vestibular Schwannoma with AICA), ***n=2/7 cerebellar hemorrhages*** 1 hemispheric and 1 midline nodulus. All had abnormal VOG and Imaging, *in one case compelling VOG findings prevented imminent discharge without imaging*.
- ***n= 4/17 Wernicke's Thiamine Deficiency*** (two had normal MRI)), n=1/17 had pontine myelinolysis
- ***n =4/17 had an Autoimmune cause*** (three paraneoplastic and n=1 post H1N1 flu ataxia). All had abnormal VOG and ***normal MRI***.
- Remaining included: 1 pontine cavernoma, 1 MS/Meniere's, 1 central with normal MRI (unknown cause), 1 post-Chiari surgery

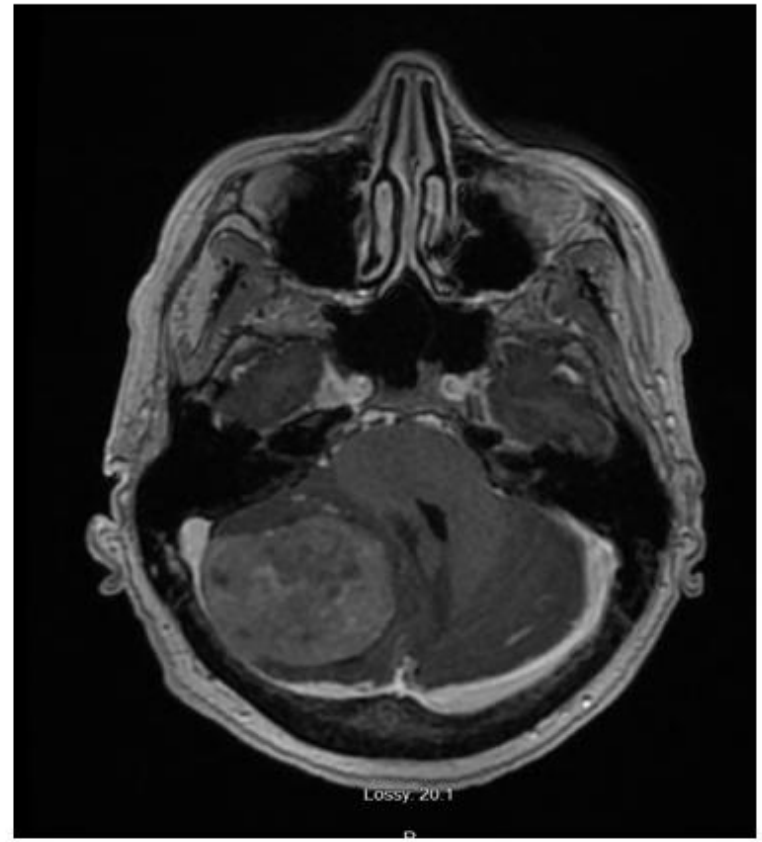
A Head Roll Left Average
Slow Phase Velocity 7 deg/sec





B Head Roll Right Average
Slow Phase Velocity 8 deg/sec





MONOPHASIC Vertigo of Unknown Cause. ABCD2 Score 3.35

- This n=10 final group had a *monophasic episode of vertigo or ataxia of unknown cause* , lasting < than 24 hours and not fitting the criteria for transient vascular vertigo , as defined by the Bárány Society.
- N=3/10 had AAS, n=7 10 AVS. VOG showed unidirectional h nystagmus in ***n=8/10*** . The vHIT showed normal gain in all. They did not have a history of migraine or Meniere's , and only one had radiographic h-CGD. One without nystagmus had no left VA signal in one single T2 MRI/MRA slice. CTA was not performed; it could perhaps fit the TIA category.

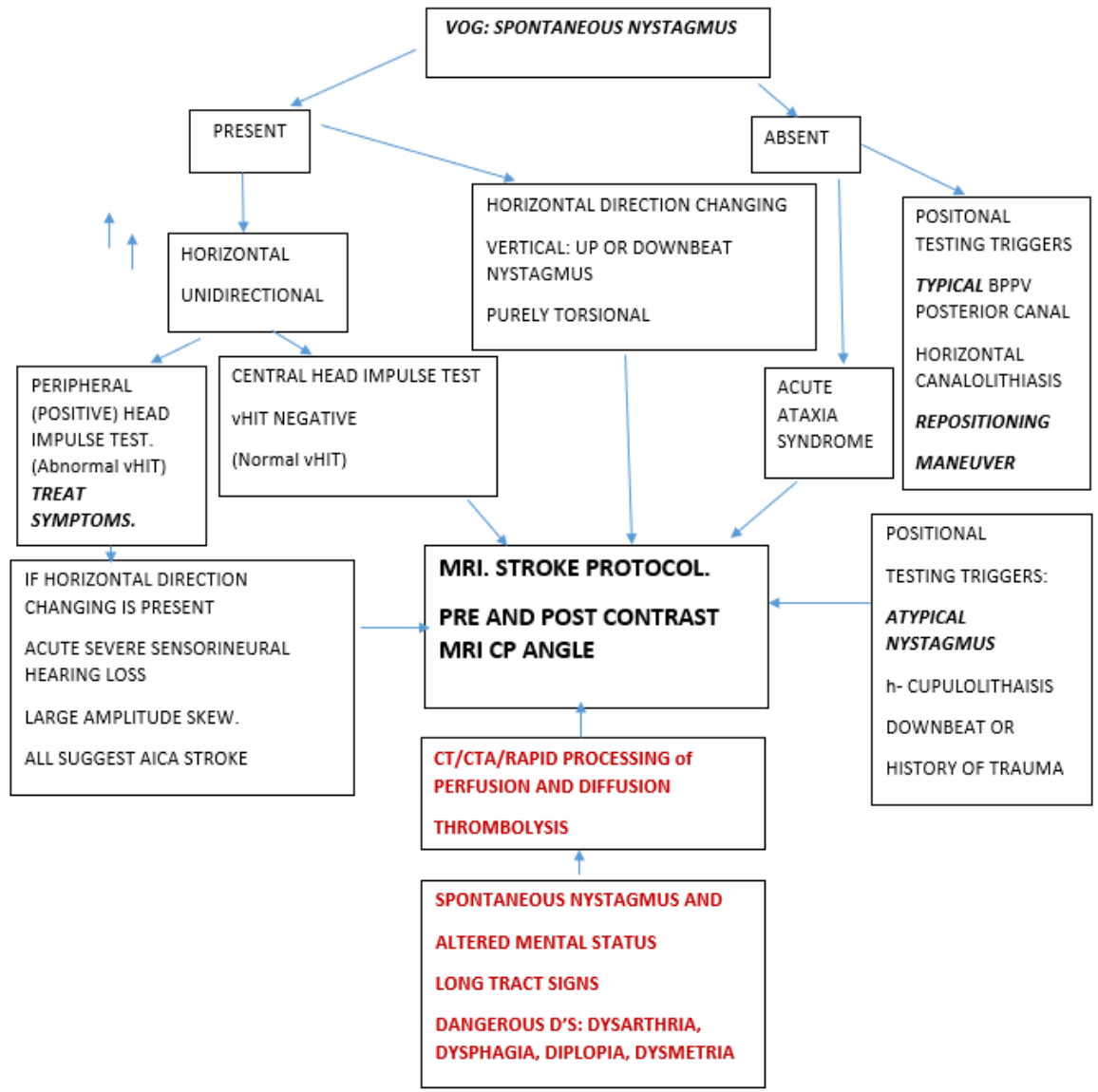
Imaging Normal in Central Cases n= 10/43 ~ 25%

- Three initially false-negative Stroke MRI
- Two Wernicke's thiamine deficiency
- Four Autoimmune or paraneoplastic
- One patient with central VOG findings , lasting longer than 24 hours without clear explanation

Unilateral Hearing Loss/Deafness, n= 7/43 ~ 16.2 %

- Three patients acute VZV
- One with chronic progressive unilateral haring loss due to vestibular schwannoma causing compression of AICA and stroke
- One with basilar artery occlusion
- One patient with inferior cerebellar peduncle and dorsal cerebellum stroke, probably labyrinthine infarct with low vHIT gain ipsilateral deafness
- One patient with severe hearing loss and posterior canal BPPV

ACUTE VESTIBULAR/ ATAXIA SYNDROME INVESTIGATION ALGORHITM



Conclusion

- Patients with AVS and AAS should undergo clinical and VOG examination. In this series, none of the patient showed rapid progression to suggest BAO; the only patient with BAO in this series had a mild AVS, with unilateral deafness and left occipital lobe micro-infarcts.
- There was time to perform the VOG which did not delay management , the majority of the patients were outside a therapeutic window
- Imaging protocol should not only address a stroke diagnosis but include pre- and post –contrast, thin sections of the CP angle to cover the differential outlined in this series.
- Radiographic h-CGD in the absence of gaze palsy in AVS/AAS should be measured and included in the imaging report.