

CLINICAL PRACTICE GUIDELINES IN NEUROTOLOGY as proposed by Dr Anirban Biswas

Clinical Practice Guidelines are as defined in the American Academy of Otolaryngology website “statements that include recommendations intended to optimize patient care that are informed by a systematic review of evidence and an assessment of the benefits and harms of alternative care options”. These are guiding principles derived by discussions and deliberations among a group of professionals in an academic / scientific association /society who are specially well versed and take special interest in a particular sub-speciality and have gathered enough clinical experience in that subject to opine on what is the best practice procedure and strategy to adopt in different clinical conditions pertaining to that particular medical discipline. Guidelines are just broad principles based on available evidences and the clinician treating the patient is expected to follow the guidelines but is not mandatory or binding on the clinician who is the best judge to decide on the management strategy as per the unique needs and requirements of the individual patient. However, there is no denying that it is always prudent and judicious to follow recommended guidelines and deviating from set principles of management as decided by experts may induce untoward results that may be deleterious to the objective of the treatment. For medico-legal reasons it is wise to document reasons for any deviations from set guidelines. The purpose of having definite guidelines for best clinical practice is to increase implementation of whatever evidence we have into practice so that clinical practice is pursued in a logical and scientific way and only those procedures and management strategies are followed which are of proven value, have stood the test of time and are based on scientific evidence as much as is possible and to avoid practices that are known to be ineffective and harmful. It is criminal to use medications that research has shown to be ineffective or deleterious to the objective of treatment. There are a lot many drugs used in neurotological disorders that have been scientifically established to be ineffective if not harmful but are yet commonly prescribed by doctors. In balance disorder patients too many biological systems are involved and the manifestation i.e., the presentation depends on the proportion of involvement of the different systems. In balance disorders it is not only the vestibular system that is involved; there is involvement of the cognitive system, the psychic system, the musculoskeletal system, the neurological system, the auditory system and sometimes also the autonomic system and the involvement of the different systems will have different effects on the patient’s physical and mental health. Each patient needs individualised and customised treatment but certain broad principles of management based on scientific evidence must be maintained and any contravention of the guidelines is expected to have disastrous outcomes due to the complexity of the problem. The guidelines are basically to ensure ethical, scientific and rational management be it diagnosis or the treatment. We must be aware that that in the truest sense no medical treatment for vestibular disorders is 100% evidence-based validated but we have to follow a consensus and accept what we have found to be logical, scientific and correct. They can serve as a guide to best practices, a framework for clinical decision making, and a benchmark. These are recommendations authenticated by the INDIAN ACADEMY OF OTOLARYNGOLOGY HEAD and NECK SURGERY (IAOHNS) and are the joint consensus document of the designated group of professionals in the IAOHNS. Available recommendations of best practice issued by other professional bodies like the European Academy of Otolaryngology and Neurotology (EAONO), the Indian Academy of Neurology, the American Academy of Otolaryngology and Head Neck Surgery and the Barany Society have been taken into account, given special weightage (due to which there are some small portions where the recommendations of some organisation like e.g., that of the Barany Society have been taken verbatim) , but re-considered and deliberated by the neurotology group in the IAOHNS to prepare the guidelines on best practice in neurotology in the Indian perspective. Though hearing disorders are very much a part of neurotology, yet other than tinnitus most hearing related issues are not included in this set of guidelines. The guidelines will be updated from time to time and more neurotological disorders will be covered in future.

Why guidelines are so important in the practice of neurotology?

Having some form of a standardised criteria for diagnosis based on consensus and clinical experience of experts is essential for disciplines like neurotology where the diagnosis is primarily symptom-driven very much like psychiatry and headache, where quite often there is no fool-proof histopathologic, radiographic, physiologic, or other independent diagnostic standard available. Though a lot of high precision vestibular function tests are available now which can pinpoint the defect in the vestibular system, yet there is substantial overlap in clinical features or biomarkers across syndromes in neurotology. The tests may show an abnormality in the cVEMP signifying a defect in the saccule of one side but the saccular defect may be the result of many different types of neurotological disorders each of which may require a completely different therapeutic approach. This does not in any way undermine the value of the vestibular function tests and rather emphasises the importance of the tests being interpreted in the right way by the right person taking into consideration different inputs obtained /extracted from the patient. In neurotology, diagnosis is based on a judicious mix of presenting symptoms, chronology of the disease, findings of the clinical tests that need to be done by the clinician, the investigative findings the most important of which are the vestibular function tests, the patient's response to previous treatment received for the disorder and above all the clinician's clinical judgement and insight in neurotology. This requires human interfacing and is not something that can be done by the computer or by remote control. Neurotology is a complete evidence based science today but the evidence requires correct clinical interpretation. The treatment is based on the aetiology of the disease, the extent of morbidity induced by the disease, the concomitant psychological and cognitive changes. Management of neurotological disorders need a holistic approach which is best possible by a trained and experienced astute clinician with special interest in neurotology.

The guidelines include the following:-

1. Specifying **personnel who are authorised to carry out the management** (investigations and/or treatment) of patients presenting with neurotological disorders (vertigo and other forms of balance disorders).
2. Specifying **the minimum clinical tests and investigations** necessary in patients presenting with vertigo and documenting them
3. Specifying **the minimum infrastructure (instruments and personnel)** of clinics where neurotological evaluation can be carried out. The minimum standards that are to be maintained in neurotological investigation reports which includes audiological tests also
4. Specifying **criteria for diagnosis and management** of different neurotological disorders
5. Specifying **nomenclature of symptoms to be used for reporting balance disorders**

1) PERSONNEL WHO ARE AUTHORISED TO CARRY OUT THE MANAGEMENT (INVESTIGATIONS AND/ OR TREATMENT)

Only medical persons specifically doctors with special interest in NEUROTOLOGY (physicians, neurologists, otolaryngologists, neurotologists) are authorised to do the vestibulometric tests and treat patients of neurotology. Even if the doctor is not doing the test himself/ herself, the vestibulometric test should essentially be done under the supervision of a qualified medical person who is adept in managing balance disorder patients and has special interest in neurotology. The tests may be done by a trained technician but only under the supervision of a medical doctor who is trained and /or has special interest in neurotology. The interpretation of the test findings in neurotological investigations is dependent on the clinical profile of the patient and needs to be analysed in the context of the clinical findings. The subject of neurotology and the balance system is unique in many ways. A normal finding in one or more of the vestibular function tests does not rule out a disorder in the balance system and a disorder found in one test may not have any bearing in the patient's balance function and may be clinically insignificant or just an incidental finding. This in no way undermines the value of the vestibulometric tests but highlights the necessity of a medical person with insight in neurotology for the relevance of the vestibulometric tests. The findings of the (correctly done) vestibular function tests have to be interpreted in the light of other findings to be clinically relevant. Neurotological diagnosis is dependent on a combination of the detailed history, the clinical findings and the results of the different neurotological tests all collated together; interpreting a neurotological report is very different from interpreting a blood report or a radiological report. As per the recommendations of the INDIAN ACADEMY OF NEUROLOGY (Ref Indian Academy of Neurology Guidelines in VERTIGO published by Elsevier 2013, page 44) and as agreed by the authorised committee making the current Practice guidelines for patients presenting with neurotological disorders, only vestibulometric reports authenticated by a doctor with special interest in vertigo should be accepted; hence medically unauthenticated reports should be summarily rejected. In vestibulometry, the quality and calibration of the instrument as well as the authenticity of the place where the tests are done are of paramount importance and the vestibulometric tests are only as reliable as the person doing the test and the clinician interpreting the test result. Faulty instruments, wrong persons performing the tests and a clinician unable to read the graphs and verify the authenticity of the reports and consequently relying on erroneous test reports (done by somebody else who is in all probability not even a medical doctor) without tallying them with clinical findings and other test results has disastrous consequences and are best abhorred. No single test is a standalone test in vestibulometry and to correctly interpret the test results a medical doctor with special interest in neurotology is essential.

2) THE MINIMUM CLINICAL TESTS AND INVESTIGATIONS NECESSARY IN PATIENTS PRESENTING WITH VERTIGO AND DOCUMENTING THEM

Clinical tests:-

In patients presenting with balance disorders first a general clinical examination that includes looking for any overt medical disorder like anaemia/pedal oedema/pulse/BP, including test for orthostatic hypotension, is mandatory. Once this is done, a basic neurological examination that includes a complete evaluation of cranial nerves (at least the third, fourth, fifth, seventh and eighth cranial nerves), tests for any motor or sensory loss in the limbs and trunk, test for planter response, tests for cerebellar function viz., finger nose tests, heel knee test and test for dysdiadokinesia and the deep tendon reflexes should be carried out. Finally, the clinical tests for balance function that comprises of the following is undertaken: –

a) VESTIBULO-SPINAL TESTS

- Standing test i.e., the Romberg's test
- Unterburger's Stepping test
- Gait test
- Walking on the floor with eyes closed and feet tandem

b) VESTIBULO-OCULAR TESTS

- Spont. nystagmus & other abnormal eye movement
- Gaze nystagmus
- Smooth tracking test
- Saccade test
- Convergence - divergence test
- Positional / Positioning tests
- Head shaking tests
- Head impulse test
- Test for skew deviation

When a patient presents with acute vertigo, the minimum clinical tests that are essentially required and the findings of which need to be documented in the clinical notes of the prescription are: –

- Test for spontaneous nystagmus.
- Head impulse test
- Test for skew deviation

○ Investigations:-

Evaluating the structural and functional integrity of the vestibular system requires a *test battery approach* i.e., a combination of different tests as no test is a standalone test as mentioned in the previous section. Also, as mentioned in the previous section, the balance system involves numerous structures and biological systems in the human body and the functional status of most if not each of these organs/systems need to be evaluated for a comprehensive assessment. Each of the structures and the involved systems has a different function which is unique in its own way and the different tests evaluate the different structures

/ systems. The three semi-circular canals monitor angular movements in three different planes, the utricle monitors front to back/ back to front and side to side movement, the saccule monitors up-down movement ; so each part of the vestibular labyrinth monitors a very specific type of movement. Testing one part does not give any information of the functionality of another part. A patient may have a defect in sensing up-down and down –up movements i.e., a saccular defect with perfect function of all other parts of the vestibular labyrinth. A VNG test or a VHIT test will be perfectly normal in such a patient and only a cervical VEMP will be able to detect this defect but this patient too will present with more or less the same complain to the doctor as a defect in any other part of the vestibular labyrinth. Hence the necessity of a test battery approach. The ENG/VNG test evaluates just the lateral semi-circular canal at a low frequency of vestibular stimulation, the oculomotor system and in many cases also helps to document presence/absence of any positional nystagmus. The oculomotor tests of VNG test only the oculomotor system, the positional tests test only for the presence of a positional vertigo and the caloric test evaluates only the lateral canal. The video head impulse test (VHIT), evaluates the status of the three semicircular canals on each side at high frequencies of vestibular stimulation. The ocular VEMP evaluates the function of the utricle and the cervical VEMP evaluates the function of the saccule. The subjective visual vertical test evaluates the perception of the visual vertical, which is a very important vestibular function. Modern research has shown the importance of evaluating the functional status of the vestibular system at different frequencies of stimulation as some diseases affect only low freq stimulation of the vestibular system and some other diseases affect only high freq stimulation of the vestibular system. The different tests of posturography like stabilometry or computerised dynamic posturography and craniocorpography evaluates the postural stability of the patient. The nerve conduction studies and the somatosensory evoked potential tests evaluate the peripheral nerves and the neural pathway involved in the maintenance of balance. A complete ophthalmological evaluation is also often necessary to rule out any defect in the visual input to the balance system. Imaging studies help to rule out any space-occupying lesion or any infarction/haemorrhage or any degenerative changes (like syringomyelia) in the parts of the brain or spinal cord that is connected with the maintenance of balance. Quite often the patient may not actually be having any balance disorder that is vertigo or imbalance at all and the patient may be having some condition like orthostatic hypotension/neuro-cardiogenic syncope/panic disorder but may presented to the doctor with the complain of head spinning only. Not only this, even many patients having primarily neurological disorders like degenerative changes affecting the cerebellum or the extrapyramidal pathways may present to the doctor complaining of instability or even head spinning. Hence all these issues have to be looked into when evaluating a patient of vertigo/imbalance. Many of the diseases causing vertigo or imbalance also have a concomitant auditory symptoms like Ménière's disease/labyrinthitis/ perilymph fistula / acoustic neuroma etc.; hence audiological tests like pure tone audiometry electro-cochleography (ECochG), brainstem evoked response audiometry (BERA) is also sometimes necessary. The astute clinician has to combine different tests to get an insight into the structural and functional integrity of the balance system and establish the exact site of lesion and the aetiology. It is not being suggested that and all patients presenting with neurotological disorders will require each and every neurotological test conceivable, but trying to establish diagnosis on the basis of one or two tests is usually not possible and the back bone of diagnosis in neurotology is a test battery approach where the right combination of different tests as thought prudent by the clinician should be undertaken. Tests should be advised liberally and not conservatively as the balance system is a very complex system and to fathom the correct diagnosis a lot of investigations are usually necessary. Whenever in doubt, it is always prudent to over investigate rather than miss diagnosis especially in patients suffering from balance disorders where the morbidity is very high and there is always more than a fair chance of a life-threatening and sinister underlying disease. However the investigations are in no way an alternative to a detailed history taking and clinical examination and the findings of the different vestibular function tests only makes sense when correlated with the history and clinical findings.

3) THE MINIMUM INFRASTRUCTURE (INSTRUMENTS AND PERSONNEL) OF CLINICS WHERE NEUROLOGICAL EVALUATION CAN BE CARRIED OUT

A neurotological clinic has the following infrastructure as regards personnel and investigative modalities i.e., instruments: –

- a. **Personnel** – a medical doctor with special interest in neurotology, qualified audiologist and trained technicians (preferably qualified computer graduates) capable of operating the diagnostic equipment. As a very detailed clinical history-taking is the first step in the approach to the management of a vertigo patient, the setup must have a dedicated person or a computer program with a proper format for detailed history taking which will be finally evaluated by the medical doctor.
- b. **Instruments** –

Vestibular function tests facilities that include:-

ENG- Electronystagmography

VNG- Videonystagmography

oVEMP - Ocular Vestibular evoked myogenic potentials

cVEMP - Cervical Vestibular evoked myogenic potentials

VHIT -Video head impulse test

DVA- Dynamic visual acuity test

SVV - Subjective visual vertical test

Posturography

CCG –Craniocorpography

the setup should also have facilities for the following: –

P T Audiometry with localising tests

BERA - Brainstem evoked response audiometry

ECochG – Electrocochleography

The **minimum requirements** should be a Videonystagmography (VNG) setup complete with oculomotor tests, a set up for video head impulse test (VHIT) and a complete evoked potential machine capable of ECochG, BERA and the ocular and cervical VEMP tests and a properly calibrated pure tone audiometry machine.

c. **Report formats:-**

1. Audiometry reports **MUST** have the last date of calibration (which is usually printed by default in most machine printouts of computerised audiometers or if a manual instrument is used then the copy of the calibration certificate) and the Masking values used during both air and bone conduction tests mentioned in the PTA report. No P T Audiometry report is complete until the masking values in dB are specified in the report
2. All evoked potential reports (BERA ECochG VEMP NCV) must have the last date of calibration of the instrument printed in the report
3. VHIT reports must mention the instrument used

4. ENG VNG BERA Tympanometry reports must have the printouts of all graphs attached with the report
5. All vestibulometric reports must be signed and authenticated by a medical doctor

4) MANAGEMENT of SOME COMMON NEUROTOLOGICAL DISEASES

A. MENIERE'S DISEASE:-

a. Criteria :

To be labelled as Meniere's disease the following criteria (as per the Barany Society, EAONO and American Academy of Otolaryngology) must be fulfilled viz

- i. Two or more spontaneous episodes of vertigo, each lasting 20 minutes to 12 hours ;
- ii. Audiometrically documented low- to medium-frequency sensorineural hearing loss in the affected ear on at least one occasion before, during or after one of the episodes of vertigo ;
- iii. Fluctuating aural symptoms (hearing, tinnitus or fullness) in the affected ear and
- iv. Not better accounted for by another vestibular diagnosis.

b. Diagnosis:

- A. The diagnosis is basically from history of episodic attacks of unprovoked vertigo that fulfil the above mentioned criteria with ear symptoms of tinnitus and deafness mainly in the low frequencies (only high freq hearing loss with normal hearing in the low frequencies is not pathognomonic or is an accepted criteria for Meniere's disease). There are criteria for definite Meniere's disease/ probable Meniere's disease / possible Meniere's disease outlined by the American Academy of Otolaryngology which may be used in qualifying the suspected Meniere's disease based on the level of evidence available for suspicion of Meniere's disease but in clinical practice the four above criteria should suffice for starting treatment of Meniere's disease

c. Investigations:

- i. The essential investigations in suspected Meniere's disease are
 1. the Pure Tone Audiometry to document the hearing loss (as without documentable sensorineural hearing loss it cannot be labelled as Meniere's disease) with localizing tests to ensure that it is a cochlear and not a neural disorder,
 2. a BERA for site of lesion to confirmatively rule out a neural lesion
 3. the Glycerol test and
 4. ECochG.
- ii. A positive glycerol test and SP:AP ratio above 0.45 in ECochG increases the index of suspicion and all patients suspected of Meniere's disease should be advised both glycerol test and ECochG even if facilities may not be available everywhere.
- iii. In diseases like Meniere's disease where a long term medication with drugs having prominent side effects are required, a very strong index of suspicion is necessary before embarking on long term treatment.
- iv. For confirmation and in doubtful cases, intra-tympanic gadolinium enhanced MRI of the inner ear of the suspected side is necessary but is not mandatory; the minimum is the typical history and the audiological tests. However such facilities are not available everywhere and even if available it need not be always done due to the costs involved and lack of standardization of technique.
- v. Vestibular Function tests at least
 1. VHIT,
 2. VNG with caloric tests and
 3. VEMP test (both cervical and ocular) with both 500Hz and 1000Hz stimulus

is indicated to document the vestibular status. The VHIT is often normal, the caloric VNG may or may not be normal and VEMP too may or may not be normal. Normal vestibular findings do not rule out Meniere's disease

- vi. Documentation of vestibular status is important for treatment and to ascertain prognosis but is not essential for diagnosis.
- vii. In early cases the caloric VNG may or may not be abnormal but VHIT is often normal.
- viii. In advanced cases where there is permanent damage of the semicircular canals of the vestibular labyrinth, the VHIT is abnormal.
- ix. In Meniere's disease if there is evidence of otolithic organ involvement (as suggested by abnormal VEMP results) the patient has chances of developing Tumarkin's crisis and is prone to sudden unprovoked falls with serious consequences and such patients should essentially be put on Meniere's prophylactic treatment even if the attacks are infrequent. VEMP should be done at both 500 and 1000Hz as a significantly higher amplitude of VEMP with a 1000Hz stimulus as compared to that with a 500Hz stimulus is very suggestive of Meniere's disease. In all other diseases and in normal ears the VEMP amplitude is usually highest with a 500Hz stimulus

d. Treatment:

- i. As regards treatment for patients having just one or less attacks in three months just a single dose of abortive treatment during the attack preferably just before the attack if the patient can predict it by noticing the aural changes or if not possible then immediately at the start of the attack with one or more of vestibular sedatives like DIMENHYDRINATE/ MECLIZINE / PROCHLORPERAZINE and a DIURETIC like furosemide and in some cases an anxiolytic drug like CLONAZEPAM / DIAZEPAM usually suffices. The patient may remain slightly sick for some hours after the attack subsides but it does not require any other treatment.
- ii. If the attacks are more than 2 attacks in three months then a long term prophylactic medication is warranted.
- iii. The *prophylactic* medical treatment is for 3 months initially and if symptoms regress both in frequency of attacks as well as in intensity then to be continued for 6 months as follows:-
 - 1. One or more DIURETICS like ACETAZOLAMIDE and SPIRONOLACTONE, FUROSEMIDE, AMELORIDE with no other medicines (i.e., vestibular sedatives) for suppression of vertigo. Dietary salt restriction to 2-4 grams /day which is often advocated is not evidenced based and is not recommended.
 - 2. If the clinician thinks it prudent or there are very frequent recurrent attacks of vertigo and /or if there are problems like electrolyte imbalance, hypotension or other problems with diuretics, then BETAHISTINE at doses above 144mg/day in 3 divided doses may be prescribed but the dosage (definitely above 144mg/day) needs to be titrated against symptoms. The pharmacology recommended dosages of 48mg/day and higher doses of upto 144mg /day has been shown to be completely ineffective and comparable to placebo in Meniere's disease. (ref, BMJ 2016;352:h6816 <http://dx.doi.org/10.1136/bmj.h6816> Christine Adrion, Caroline Simone Fisher, Michael Strupp et al; Efficacy and Safety of Betahistine treatment in patients with Meniere's disease: primary results of a long term , multicenter, double blind, randomized, placebo controlled dose defining trial (BEMED trial) ; British Medical Journal 2016:352). Hence the doses of betahistine below 144mg/day are not recommended
 - 3. Depending on severity of symptoms the clinician may in the initial stages use both diuretics as well as betahistine (above 144mg/day) together.
- iv. Avoidance of Nicotine and caffeine especially caffeine-containing food and drinks, such as coffee, tea, and chocolate, is recommended alongwith.
- v. If this treatment fails (no significant reduction in intensity and frequency of attacks of vertigo) then and only then Intratympanic Gentamicin(ITG) low-dosage (26.6mg/ml) treatment is recommended after a trial of at least 6 months of medical treatment. ITG treatment is to be

attempted at a frequency not more than once every 7 days and the reduction (if any) in symptoms monitored.

- vi. If no significant improvement takes place then after 3 injections the ITG treatment should be stopped and the patient observed for at least 3 months.
- vii. After that a decision on Surgical treatment in the form of selective Vestibular neurectomy (in case of serviceable i.e., residual hearing better than 70dB is present) but if there is no serviceable hearing then a Labyrinthectomy is to be done. If surgical treatment (i.e., labyrinthine de-afferentation) is done then rigorous and diligent VESTIBULAR REHABILITATION exercises are mandatory for the first 6 weeks after surgery.
- viii. In bilateral Meniere's disease, autoimmune background should be considered; hearing loss is generally the major concern, choice of treatment (especially in failure on medical treatment) should be individualized but Steroids should be included but otherwise treatment should be in the same lines as unilateral Meniere's disease except that ITG treatment and labyrinthectomy should best be avoided as there is a definite chance of further hearing loss in these procedures especially in the later.

B. BPPV: -

- a. to be suspected from the typical history of very brief spells of vertigo only on change of head position and diagnosed confirmatively only by clinical tests ;
- b. If required a Video Frenzel glasses may be used for documentation but clinical naked eye examination of the patient's eyes is sufficient to diagnose any positional vertigo.
- c. Ordinarily the detailed vestibular function tests (i.e., investigations of vestibular function like VNG, VHIT, VEMP, SVV, DVA, posturography) are not recommended if history and clinical positional tests like Dix Hallpike tests are typical of BPPV.
- d. The first line treatment in posterior canal BPPV is either Epley's or Semont's maneuvers, not both together.
- e. Medical treatment with antivertigo drugs have NO role in the treatment of definite posterior canal geotropic benign positional vertigo which is the commonest form of positional vertigo and even if the positional vertigo is ageotropic.
- f. Mastoid vibration is not recommended during the maneuvers as they have not been found to be additionally beneficial.
- g. Vestibular sedative medications (antivertigo drugs) like Prochlorperazine or Diazepam may be used as a single dose only to improve compliance during maneuvers.
- h. In case of failure of first maneuver the other maneuver may be tried or the same maneuver repeated. But if the first maneuver fails, before trying a maneuver once again it is judicious to re-think about the diagnosis and repeat the detailed history taking as well as the clinical neurotological assessment of the patient and carry out a detailed vestibular function test.
- i. Just one maneuver if properly done suffices in one session and repeating the same maneuver several times is best avoided.
- j. The Brandt-Daroff exercises at home are not to be advised if the Epley's or Semont's maneuver is properly and successfully done. The Brandt-Daroff exercises at home are best recommended for those patients who have a very mild positional vertigo or on cases where the symptom of vertigo only without any perceptible nystagmus is there during the positional tests or if the clinician is not too sure about the side of the positional vertigo in Dix Hallpike tests.
- k. Surgery (canal plugging or singular neurectomy) is not recommended before 1 year of follow-up and is to be done only after the maneuvers have repeatedly failed but the diagnosis of benign positional vertigo of the posterior canal is very certain.
- l. Post maneuver movement restrictions are not recommended. There is no restrictions to driving a few hours after the maneuver is done.
- m. In geotropic lateral canal BPPV which is diagnosed by the side roll test by making the patient turn the head laterally to the left / right in straight supine position first line treatment is the

Barbecue, Gufoni or Vannucchi maneuvers. Just one of the three maneuvers not all three together is recommended.

- n. Like the posterior canal BPPV vestibular sedative medications (antivertigo drugs) have a role only to improve compliance during maneuvers and may be given just once prior to the maneuver in very apprehensive patients but there is NO role of anti vertigo drugs in lateral canal BPPV.
- o. If one maneuver is ineffective then the same maneuver may be repeated or another maneuver i.e., one of the two others may be tried. If the first maneuver fails in spite of doing it satisfactorilya rethink on the diagnosis and a detailed vestibular function test is recommended. Here too mastoid vibration and post maneuver restriction of head movement is not recommended.
- p. Surgical plugging of the lateral canal is recommended after at least one year of repeatedly trying the recommened maneuvers and repeatedly failing to provide relief for a substantial period with the properly carried out maneuvers.
- q. In ageotropic lateral canal BPPV, try to transform the ageotropic nystagmus to geotropic to exclude central vestibular system involvement. This is often possible by vigorous head shaking. After that Gufoni/modified Gufoni or Vannucchi maneuvers is to be tried. Whenever one maneuver is found ineffective another maneuver for the same BPPV typee is to be tried or a repetition of the first maneuver may be attempted but before that thae patient must be thoroughly reassessed for confirmation of diagnosis and to exclude any other possible cause of vertigo.
- r. Anterior canal BPPV is a controversial entity and there is currently no consensus on treatment though many maneuvers have been proposed. A differentiation from a cerebellar disorder is to be carried out whenever anterior canal BPPV is suspected.
- s. Whenever there is a suspecion of multiple canal involvement one canal at a time is to be tried and the most symptomatic canal should be tried first by the requisite maneuver.
- t. Even in multiple canal BPPV there is no scope of continued medical treatment with anti vertigo drugs. Just one dose if required half an hour before the manuever in very apprehensive patients is the maximum permissible.

C. VESTIBULAR NEURONITIS: -

1. A better term is Acute Unilateral Vestibulopathy as the clinician is not sure about the pathophysiology when the patient presents.
2. Presentation:
 - i. The presentation is usually of an acute onset of sudden severe vertigo which often persists for more than one day but definitely less than seven days the usual period being one to three days.
 - ii. There is no accompanying deafness or any other aural symptoms and no CNS symptoms like headache, diplopia, any motor/sensory loss or any drowsiness or loss of consciousness.
 - iii. Classical features like this are not usually present as most patients have some vestibular sedatives or CNS depressants or some anti-emetics (because of the accompanying nausea-vomiting along with the vertigo) which alters the presenting feature.
 - iv. The classical clinical signs are a direction fixed horizontal nystagmus beating either to the left or to the right without any skew deviation of the eyes and a positive head impulse test on the side opposite to the direction of nystagmus (i.e., a left sided head impulse test +ve with a right beating nystagmus).
 - v. Any ataxia must always be looked for in vestibular neuronitis the patient should not at all be ataxic though a mild instability is not unusual. Ataxia or severe instability is strongly suggestive of a central lesion. In all patients presenting with sudden onset acute vertigo a cerebellar stroke must be mandatorily ruled out as acute cerebrovascular accident in the cerebellum often presents with a perfectly similar

presentation and may perfectly mimic vestibular neuronitis. The most effective way of ruling it out within the first 24 hours is the HINTS test.

1. The clinician must specifically carry out

- A. the head impulse test clinically or better still if available then a VHIT test; - a +ve head impulse test rules out a cerebellar stroke and confirms that the nystagmus is due to a peripheral vestibulopathy but a negative head impulse test strongly suggests that there is a CVA involving the cerebellum ,
 - B. look at the nystagmus at least for 2-3 minutes and see whether the direction of the horizontal nystagmus is fixed i.e, always to one direction or is it changing in direction i.e., a left beating nystagmus becoming right beating and vice versa, the clinician should also specifically look for any vertical nystagmus - any direction changing nystagmus or any vertical nystagmus is pathognomonic of a central lesion like a cerebellar stroke in such patients and
 - C. The clinician must also specifically look for any skew deviation of the eyes or any ocular tilt reaction - any skew deviation or any abnormal ocular tilt suggests a central lesion.
- vi. The differential diagnosis to be considered in such cases where the patient presents with acute vertigo are
1. first attack of an attack of Vestibular Migraine ,
 2. first attack of Meniere's disease where the aural symptoms may not be present or are so mild that they are not noticed by the patient and what is most important is
 3. A cerebellar / brainstem stroke.
- vii. The diagnosis is primarily by a thorough history and a clinical examination where at least the clinical tests mentioned above are carried out and documented.
- viii. However if there is any sign of a central lesion like -ve head impulse test, direction changing nystagmus or vertical nystagmus, skew deviation or definite ataxia then an urgent MRI of the brain esp focus on the cerebellum and brainstem is suggested and if the MRI is normal then a repeat MRI with contrast after 24 hours of onset is suggested.
- ix. But if the central features are not there on clinical examination then the MRI of brain is not indicated.
- x. Treatment of vestibular neuronitis i.e., if a central lesion has been ruled out then;-
1. first line treatment is symptomatic treatment with prochlorperazine for first 1-3 days only and steroids (I.V. steroids for 1-2 days if patient is vomiting) otherwise oral steroids which is slowly tapered off.
 2. The patient is encouraged to be active and to move about, stay in a lighted room and to start the vestibular rehab exercises from day one initially only the exercises in sitting position and then as soon as possible the standing and walking exercises.
 3. If some symptoms persist even after seven days then a fresh neurotological assessment of the patient and a re-think on the diagnosis and the detailed vestibular function tests that include VNG, VHIT, ocular and cervical VEMP, dynamic visual acuity, subjective visual vertical test etc along with a pure tone audiometry are to be carried out.
 4. In all such patients rigorous Vestibular rehabilitation exercises is mandatory and the clinician should instruct the patient and family members / care givers accordingly.
 5. There is no indication for surgical treatment in all such patients.
 6. In case of long-term residual dizziness after unilateral vestibular loss, no medical treatment is recommended and in all such patients' proper counselling and rigorous vestibular rehab exercises is recommended.

7. Of course in case the detailed vestibular function tests have not been done then all the vestibular function tests mentioned above are to be essentially done along with imaging studies (MRI) of the brain.
8. Vestibular sedatives / anti-vertigo drugs should never be continued in all such patients for more than 3-7 days at the most. The consensus is to limit it to 3-5 days only and this is the recommendation of the IAOHNS.
9. In fact many authorities suggest that drugs that exert a sedative effect on the vestibular system should be discontinued after the first 24 hours.

D. **Migraine Related Vertigo/ Migraine Associated Vertigo /Vestibular Migraine**

- a. **History:** History is the most important means to diagnose migraine-associated vertigo. Patients with migraine-related vestibulopathy typically experience a varied range of dizzy symptoms throughout their life and the attacks are not necessarily the same every time; attacks may be simple head spinning i.e., a spinning/ rotating sensation (true vertigo) or may be phases of just ataxia/ imbalance without any true head spinning or may even be phases of light headedness or may be a combination of vertigo, light-headedness and imbalance. A thorough headache history is also important when evaluating patients for possible migraine-associated vertigo. No definitive diagnostic tests exist which is pathognomonic for migraine-associated vertigo. When the history is unclear, the diagnosis is made by a therapeutic response to treatment and by ruling out other possible causes. Due to its protean manifestations and varied nature of presentation as well as the lack of any marker / definite diagnostic test, diagnosis is difficult and Vestibular Migraine is basically a diagnosis by elimination. In a suspected case of Vestibular Migraine due to lack of definitive diagnostic criteria requisite notes of history and clinical findings must be documented in the prescription to avoid ambiguity later. A family history of Migraine, history of motion sickness and history of typical Migraine headaches either time-locked with the vertigo or even if occurring separately are important diagnostic parameters and the presence / absence of any of these should be specifically asked for and documented.

Diagnostic criteria for Vestibular migraine as proposed by the Barany Society and other organisations like International Headache Society with small changes (numbers in superscript detailed below) are as follows:-

1) **Criteria for *Definite Vestibular Migraine***

- A. At least 5 episodes with vestibular symptoms¹ of moderate or severe intensity², lasting 5 min to 72 hours³
- B. Current or previous history of migraine with or without aura according to the International Classification of Headache Disorders (ICHD)⁴
- C. One or more migraine features with at least 50% of the vestibular episodes⁵:
 - headache with at least two of the following characteristics:
 - One sided location, pulsating quality,
 - Moderate or severe pain intensity, aggravation by routine physical activity
 - photophobia and phonophobia⁶,
 - visual aura⁷
- D. Not better accounted for by another vestibular or ICHD diagnosis⁸

2) **Criteria for *Probable vestibular migraine***

- A. At least 5 episodes with vestibular symptoms¹ of moderate or severe intensity², lasting 5 min to 72 hours³
- B. Only one of the criteria B and C for vestibular migraine is fulfilled (migraine history or migraine features during the episode)
- C. Not better accounted for by another vestibular or ICHD diagnosis⁸

Notes:

1. Vestibular symptoms include:

- spontaneous vertigo including
 - internal vertigo, a false sensation of self motion, and
 - external vertigo, a false sensation that the visual surrounding is spinning or flowing,
- positional vertigo, occurring after a change of head position,
- visually-induced vertigo, triggered by a complex or large moving visual stimulus
- head motion-induced vertigo, occurring during head motion,
- head motion-induced dizziness with nausea.

Dizziness is characterized by a sensation of disturbed spatial orientation. Other forms of dizziness are currently not included in the classification of vestibular migraine.

- ❖ Vestibular symptoms are rated “moderate” when they interfere with but do not prohibit daily activities and “severe” if daily activities cannot be continued.
- ❖ Duration of episodes is highly variable: About 30% of patients have episodes lasting minutes, 30% have attacks for hours and another 30% have attacks over several days. The remaining 10% have attacks lasting seconds only, which tend to occur repeatedly during head motion, visual stimulation, or after changes of head position but sometimes even completely unprovoked. In these patients, episode duration is defined as the total period during which one single attack of vertigo / dizziness / imbalance persists. At the other end of the spectrum, there are patients who may take a few weeks to fully recover from an episode. However, the core episode rarely exceeds 72 hours. However though as per most guidelines attacks of vestibular migraine should not last more than 72 hours
- ❖ Migraine categories 1.1 and 1.2 of the ICDH[International Headache Society Classification Subcommittee, International Classification of Headache Disorders. 2nd Edition, *Cephalalgia*24(Suppl 1) (2004)].
 - **1.1 Migraine without aura:** Previously used terms: Common migraine; hemicrania simplex. Description: Recurrent headache disorder manifesting in attacks lasting 4-72 hours. Typical characteristics of the headache are unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity and association with nausea and/or photophobia and phonophobia. Diagnostic criteria: A. At least five attacks fulfilling criteria B–D B. Headache attacks lasting 4-72 hours (untreated or unsuccessfully treated) C. Headache has at least two of the following four characteristics: 1. unilateral location .2 pulsating quality. 3 moderate or severe pain intensity 4. aggravation by or causing avoidance of routine physical activity (e.g. walking or climbing stairs) D. During headache at least one of the following: 1. nausea and/or vomiting 2. photophobia and phonophobia E. Not better accounted for by another ICHD-3 diagnosis.
 - **1.2 Migraine with aura:** Recurrent attacks, lasting minutes, of unilateral fully reversible visual, sensory or other central nervous system symptoms that usually develop gradually and are usually followed by headache and associated migraine symptoms. Diagnostic criteria: A. At least two attacks fulfilling criteria B and C B. One or more of the following fully reversible aura symptoms: 1. visual 2. sensory 3. speech and/or language 4. motor 5.

brainstem 6. retinal C. At least two of the following four characteristics: 1. at least one aura symptom spreads gradually over 5 minutes, and/or two or more symptoms occur in succession 2. each individual aura symptom lasts 5-60 minutes 3. at least one aura symptom is unilateral 4. the aura is accompanied, or followed within 60 minutes, by headache D. Not better accounted for by another ICHD-3 diagnosis, and transient ischaemic attack has been excluded.

- ❖ One accompanying symptom is sufficient during a single episode. Different symptoms may occur during different episodes. Associated symptoms may occur before, during or after the vestibular symptoms.
- ❖ Phonophobia is defined as sound-induced discomfort. It is a transient and bilateral phenomenon that must be differentiated from recruitment, which is often unilateral and persistent. Recruitment leads to an enhanced perception and often distortion of loud sounds in an ear with decreased hearing.
- ❖ Visual auras are characterized by bright scintillating lights or zigzag lines, often with a scotoma that interferes with reading. Visual auras typically expand over 5–20 minutes and last for less than 60 minutes. They are often, but not always restricted to one hemi-field. Other types of migraine aura, e.g. somatosensory or dysphasic aura, are not included as diagnostic criteria
- ❖ History and physical examinations do not suggest another vestibular disorder *or* such a disorder is considered but ruled out by appropriate investigations *or* such disorder is present as a comorbid or independent condition, but episodes can be clearly differentiated. Migraine attacks may be induced by vestibular stimulation. Therefore, the differential diagnosis should include other vestibular disorders complicated by superimposed migraine attacks.

There is also a class of patients who have recurrent episodes of true vertigo / instability without any other accompanying symptoms related either to the ears or to the central nervous system and do not have any history of headaches. The duration may be anything between a few secs to more than 72 hours. The important criteria are the recurrent episodic nature of the presentation with perfectly normal symptom free periods. Though this does not fall directly into the Barany Society or the IHS criteria, yet there are a large number of patients who fall in this category. These patients may be diagnosed as *Possible Vestibular Migraine* and warrants a therapeutic trial with migraine prophylactic therapy. The very good response obtained from most of these patients with migraine therapy justifies the therapeutic trial provided of course other causes of vertigo have been ruled out.

Transient auditory symptoms, nausea, vomiting, prostration, and susceptibility to motion sickness maybe associated with vestibular migraine. However, as they also occur with various other vestibular disorders they are not included as diagnostic criteria. Provocation of an episode can be a diagnostic clue. Menstruation, stress, lack of sleep, dehydration, and certain foods that are known to trigger migraine headaches may all trigger attacks of Migraine associated Vertigo / Vestibular migraine, but are not included as diagnostic criteria. However the history of such precipitating factors must be asked for and documented.

The main differential diagnosis is from:-

- Meniere's Disease
- BPPV
- Episodic Ataxia type 2
- Spontaneous Mal de debarquement syndrome
- Transient Ischemic Attacks

Requisite leading questions to rule out these clinical conditions need to be asked to every patient suspected of having Vestibular Migraine. The salient features of these abovementioned disorders is beyond the scope of this guideline.

The most difficult differentiation of Vestibular Migraine is from Meniere's disease and positional vertigo (BPPV).

a. Physical examination:

Complete neurotologic and Neurologic clinical examination Neurotologic examination are often normal. Horizontal rotary spontaneous nystagmus may be present during an acute attack of vertigo be it Vestibular Migraine or BPPV but in BPPV it will only be in specific positions of the head and for very brief durations. There will be no nystagmus in other head positions or when the patient is sitting straight or lying flat. Dix-Hallpike examination may elicit symptoms of vertigo with or without nystagmus. When it is without nystagmus and in multiple head positions the first diagnosis should be Vestibular Migraine as migraine patient patients very often have motion intolerance and sudden change of head position elicits a severe discomfort which they report as vertigo; however presence of nystagmus in different head positions does not rule out Vestibular Migraine. Vestibular migraine may present with purely positional vertigo, thus mimicking BPPV; hence differentiation is important. In vestibular migraine, positional nystagmus if at all present is usually persistent, occurs in multiple head positions and not aligned with a single semicircular canal. Very often there is no nystagmus but patient complains of vertigo in vestibular migraine because of the motion intolerance as explained above. Symptomatic episodes tend to be shorter with vestibular migraine (minutes to days rather than weeks) and more frequent (several times per year with vestibular migraine rather than once every few months / years with BPPV). In BPPV there is definite nystagmus in only one or two specific head positions and the history is of vertigo of brief durations (secs only) that occurs recurrently for a few days at a stretch whereas in Vestibular migraine there may be one attack in a few days or a few months and usually not over a span of several days followed by a symptom free period of several months which is typical of BPPV. When in doubt it is wise to try a suitable liberatory maneuver like Epley's / Semont's/ Guffoni's etc; BPPV should respond very well to the liberatory maneuvers but Vertiginous Migraine will not; in fact if a patient presenting with typical positional vertigo does not respond adequately to correctly done liberatory maneuvers Vestibular Migraine should be thought of as the cause and a therapeutic trial with Migraine prophylactic medication instituted. There is a complicated relationship between Vestibular Migraine and BPPV. BPPV patients are more likely to have motion sickness and / or Migraine headaches. Patients with subjective positional vertigo but without typical signs are more likely to have migraine (ref Agarwal, Bronstein 'Visual Dependence and BPPV' journal of Neurology 2011;259:1117-24) It is however possible that as migraine patients are usually very sensitive to motion they probably seek medical attention more easily than non migraine patients. Vertigo attacks are known to trigger migraine. The relationship is complex and because of the overlap of symptoms the differentiation between positional vertigo and vestibular migraine is often not very easy but essential in all patients of suspected vestibular migraine.

Fluctuating hearing loss, tinnitus and aural pressure may occur in vestibular migraine, but hearing loss does not progress to profound levels as in Meniere's disease

b. Investigations :

Investigations suggested are (1)PT audiometry (even if the patient does not complain of any hearing disorder), (2)Vestibular Function tests at least a VNG with oculomotor tests and caloric test, cervical and ocular VEMP at 500 and 1000Hz, Video Head Impulse Test and (3) Imaging of the brain

1. **No pathognomonic abnormalities on imaging studies or vestibular testing** either confirm or rule out migraine-associated vertigo. They are helpful only to rule out other causes of vertigo.

2. **Audiometric evaluation**

Pure-tone audiometry, word recognition scores, and reflex testing –is usually normal. Audiometry may show concurrent hearing loss which may be due to other causes or sometimes due to migraine induced spasm of the cochlear blood supply; hence though deafness is not expected in Migraine associated vertigo, yet presence of deafness /ear symptoms does not rule out migraine associated vertigo. But in such cases where deafness is present, differentiation from Meniere's disease is difficult by history alone. When the criteria for Meniere's disease are met, particularly hearing loss as documented by audiometry, Meniere's disease should be the first diagnosis, even if migraine symptoms occur during the vestibular attacks. Under such circumstances where there is a diagnostic dilemma, investigations like multi-frequency VEMPs, ECochG and Glycerol tests are helpful as there are definite diagnostic features for all these investigations in Meniere's disease

3. **Electronystagmography(ENG) or videonystagmography (VNG)** is typically not helpful in differentiating migraine-associated vertigo from Ménière disease. However, for patients with a several-year history of dizziness, normal findings on ENG are suggestive but never diagnostic of migraine-associated vertigo. Documentable oculomotor problems are suggestive of Migraine associated Vertigo

4. **Electrocochleography**

Patients with a several-year history of Ménière disease often have a reduced vestibular response on at least 1 side. Electrocochleography (ECochG) may help to differentiate Ménière disease from migraine-associated vertigo. A high SP:AP ratio is indicative of Meniere's disease

5. **Vestibular evoked myogenic potential**

Cervical vestibular evoked myogenic potentials (cVEMPs) : In vestibular migraine and even normal ears the amplitude of cVEMP is higher if 500Hz stimulus is used compared to 1000 hz stimulus. In Meniere's disease the cVEMP amplitude with 1000Hz is higher than with 500Hz stimulus. This is a diagnostic feature of Meniere's disease and helps to rule out Migraine Associated Vertigo

6. **Caloric testing** : Caloric unilateral or bilateral weakness may be seen but is not diagnostic. Calorization may in fact trigger an attack of Migraine associated Vertigo.
7. MRI of the brain with gadolinium is necessary when patients present with unilateral symptoms or signs or if the patient's symptoms do not respond to appropriate treatment. If the patient's symptoms are those of unilateral sensorineural hearing loss or tinnitus along with vestibular symptoms, the MRI is indicated with special focus to the internal auditory canals, brain stem and cerebellum. There is no diagnostic feature in brain MRI that is pathognomonic of Vestibular Migraine.

C. Treatment :

Dizziness secondary to migraine usually responds to the same treatment used for migraine headaches. The choice of drugs is mainly guided by the frequency of the attacks and the side effect profile.

The 3 broad classes of migraine headache treatment include reduction of risk factors, abortive medications, and prophylactic medical therapy

1. Reduction of risk factors includes an attempt to avoid certain conditions (eg, stress, anxiety, hypoglycemia, fluctuating estrogen, certain foods and smoking) that can trigger migraine. Elimination of birth control pills or estrogen replacement products. The following foods should be avoided:
 - Monosodium glutamate (MSG)
 - Certain alcoholic beverages - eg, red wine, port, sherry, bourbon
 - Aged cheese , fermented food
 - Chocolate

- Aspartame

An elimination diet for 6 weeks minimum may be prescribed. If, after 6 weeks, symptoms are not better, diet modification only is not considered to be helpful. If foods are a trigger for symptoms, the offending food(s) can be identified by adding back 1 food at a time until the symptoms return.

B. Abortive medication and risk-factor reduction

In general, drugs used to abort migraine headaches like the triptans have not been found effective in treating dizziness secondary to migraine (i.e., aborting an acute episode of Migraine induced vertigo).

C. Prophylactic pharmacotherapy

- First-line prophylactic medications (drugs established as effective) : Divalproex Sodium (400-1000mg), topiramate (50 to 150mg/day), Sodium Valproate, metoprolol (47.5 to 200mg/day), propranolol (80 to 160mg/day), venlafaxine (37.5 to 75mg/day), and methysergide..
- Second-line treatments (drugs probably effective): calcium channel blockers (verapamil, flunarizine), tricyclic antidepressants (nortriptyline, amitriptyline (25-100mg/day)), riboflavin, atenolol.

Betablockers (metoprolol and propranolol) induce low BP, fatigue, constipation bradycardia, asthma, possibly mental depression in some patients; Tricyclic antidepressants induce tachycardia, tachy-arrhythmia, and urinary retention. Topiramite causes wt. loss, paraesthesia, speech problems and difficulty with memory and may aggravate narrow angle glaucoma and cause depression and personality change. Sodium Valproate induces wt. gain, and also causes sedation, GI-upset, rarely tremor and very rarely hepatic damage. Flunarizine induces weight gain and depression and in addition has all the negativities of a Ca channel blocker and causes pedal edema, Parkinsonism etc

Acetazolamide and lamotrigine are also recommended drugs primarily effective for only the vestibular symptoms and not headaches

If dizziness is not controlled with one class of medication, another class should be used. If dizziness is controlled with one of these medications, the drug should be administered continuously for at least 1 year (except for methysergide, which requires a 3- to 4-week drug-free interval at 6mo). The medication can be restarted for another year if the dizziness returns after discontinuing therapy.

Referral to vestibular rehabilitation should be considered for all patients, particularly if secondary complications such as deconditioning, loss of confidence in balance or visual dependence have developed.

E. VESTIBULAR PAROXYSMIA practice guidelines

Vestibular paroxysmia (VP) previously known as disabling positional vertigo or vascular loop syndrome is a controversial, vestibular disorder/syndrome caused by neurovascular cross compression of the vestibular nerve with features of recurrent, spontaneous, episodic very brief attacks of subjective / objective vertigo or even instability that generally last less than one minute (often a second or even lesser) but may rarely be for a few minutes also and occur in a series of clusters of repeated identical attacks. If the attack is that of spinning in a particular direction it is always just spinning in that direction or if it is a momentary instability or a feeling of later propulsion (being pushed) it is always exactly the same for the particular patient. Consistency of the nature of symptoms is one of the typical features of this difficult to diagnose clinical entity. The number of attacks is anything between numerous every day to a few discrete attacks a month. The single discrete attacks subside by

themselves without intervention but keeps on repeating. The attacks of vertigo/ instability are sometimes but not always accompanied with auditory symptoms which may occur either just during the attack of vertigo or sometimes be there even when the vertigo is not present. Positive response to carbamazepine therapy strongly suggests but does not prove that the vestibular disorder is due to vestibular paroxysmia. When present with auditory symptoms (esp. tinnitus) the condition is also referred as audio-vestibular paroxysmia. During the attack there is usually some documentable neurophysiologic change in the audio-vestibular tests. Diagnosis by investigations only is problematic as (1) detectable audio-vestibular abnormalities may or may not be present i.e., though some abnormalities are often found yet in many patients audio-vestibular findings are perfectly normal; (2) MRI has very high sensitivity but poor specificity i.e., radiological evidence of a very close vascular loop on the 8th cranial nerve is present in a third of normal patients and so just the presence of a vascular loop in very close proximity of the vestibular nerve does not mean conform vestibular paroxysmia; and (3) side determination is very difficult if not impossible esp in patients who have only vestibular symptoms. Diagnosis is hence by a combination of clinical features (i.e., history), audio vestibular tests and specialised MRI studies and of course the clinician's strong index of suspicion. .

PATHOPHYSIOLOGY and FEATURES:

A blood vessel, mostly the anterior inferior cerebellar artery(AICA) or sometimes the posterior inferior cerebellar artery(PICA), vertebral artery or vein, vascular malformation, megalodolichobasilar artery, etc. which forms a loop around the vestibular branch of the cochleo-vestibular nerve (VIII cranial nerve) in the area proximal to the "transition zone" is responsible for this syndrome, similar to the other neurovascular cross compression syndromes like hemi-facial spasm and trigeminal neuralgia. The site of lesion is the root-entry-zone of the 8th cranial nerve where myelination is by Schwann cells. This neurovascular loop formation causes pressure on the nerve along with cross-compression resulting in lesion with demyelination of the central myelin, proximal to the "transition zone" i.e. first 1.5 cm after the nerve exit. Consequently, short circuits (erratic stimulation) occur in the nerve conduction resulting in intermittent nerve block which is responsible for the brief attacks of spinning vertigo (quick spins), postural vertigo and/or instability. The attacks may also be accompanied by unilateral hearing loss and tinnitus if neighbouring cochlear branch is involved. Triggering factors or worsening factors for these attacks may not be there but when present include certain head position(s), hyperventilation and include change in head movements or certain exercises.

Diagnosis: – as in most other neurotological disorders, diagnostic criteria for vestibular paroxysmia have been specified for (a) definite, and (b) probable vestibular paroxysmia. Since there are some grey areas in our understanding of the exact pathophysiology and due to the protean manifestations of the disease very much like Vestibular migraine, different schools have proposed slightly different criteria for labelling a disorder as Vestibular Paroxysmia. The author(s) after examining all different guidelines and diagnostic parameters proposed by different authors and groups have thought it prudent to follow the proposals of the Barany Society with minor modifications.

To be labelled as **definite vestibular paroxysmia** the following criteria have to be met:-

1. there must be at least 10 attacks of either subjective or objective spinning or instability each for less than one minute
2. should occur spontaneously that is completely unprovoked
3. each of the attacks be similar in intensity and quality that is stereotyped
4. should respond to a therapeutic trial with Carbamazepin or oxcarbazepine
5. not better accountable by other causes of vertigo

To be labelled as **probable vestibular paroxysmia** the following criteria have to be fulfilled: –

1. at least five attacks of subjective or objective head is spinning none of them being for a duration of above five minutes

2. attacks may be spontaneous, that is unprovoked or maybe precipitated in certain head positions but does not respond to corrective liberatory manoeuvres and are not in the typical Dix Hallpike or other BPPV specific positions
3. each attack in an individual is similar in intensity and quality that is the vestibular symptoms are stereotyped
4. not better accountable by other causes of vertigo

Differential diagnosis: – as vestibular paroxysmia is not one of the commonest cause of vertigo, the following clinical conditions (esp the first three) which are more common but may have a more or less similar presentations need to be ruled out. In general, the diagnosis has to be very strongly suspected on the basis of history /clinical examination/investigations before venturing on a therapeutic trial with Carbamazepin/oxcarbazepine which have of dangerous adverse effects

1. vestibular migraine
2. vestibular epilepsy/ vestibular seizures
3. BPPV
4. transient ischaemic attacks of the posterior circulation
5. perilymph fistula (the trigger factor is very definite like increased intra-abdominal pressure, sneezing, coughing, lifting heavy weights etc)
6. superior semi-circular canal dehiscence
7. panic attacks
8. atypical brainstem events as in some cases of multiple sclerosis
9. Tumarkin's crisis provided a typical history of Meniere's disease is present
10. If it occurs only on turning or rotating the head to one side and the feeling is more of a sinking / falling/ blurring and not typically the spinning sensation then occlusion of the vertebral artery is a remote possibility and should be looked for by requisite imaging tests like angiography. If confirmed, then the diagnosis is Rotational Occlusion of Vertebral Artery Syndrome

Clinical diagnosis is no doubt difficult as this disease does not have any definite marker either on clinical tests or on investigations which are pathognomonic of vestibular paroxysmia. The clinician should understand that the mere MRI finding of vascular compression of the proximal part of the eighth cranial nerve is not a sure shot evidence of clinical vestibular paroxysmia as about one third of normal subjects also have similar radiological findings without any symptoms of vertigo whatsoever. Positive therapeutic response to Carbamazepin/oxcarbazepine is very suggestive but not a fool proof evidence of vestibular paroxysmia as there is not enough of supportive randomised, double-blind, placebo-controlled multicentre studies are available in clinical literature till now.

Investigations: – The audio-vestibular tests will show deficits during the periods of attacks which are less noticeable during asymptomatic intervals when there are no attacks. In symptom-free periods there may not be any detectable abnormality in the audio-vestibular tests and normal findings in these tests do not rule out vestibular paroxysmia. The tests that can be conducted include audiogram, BERA, VNG with oculomotor and caloric tests, VHIT, VEMP, SVV. Prolongation of I-III inter-peak latency can sometimes be found in BERA but only if the auditory nerve has also been involved where there will also be some auditory symptoms but otherwise the BERA is expected to be normal. The VNG caloric weakness (canal paresis above 20%) and spontaneous nystagmus (time-locked to the attack and beating toward the affected ear) may be noted in caloric tests. Low amplitude or delayed wave peaks in cervical and /or ocular VEMP are also possible but none of the tests have specific role in confirming or even in diagnosing Vestibular Paroxysmia. Nevertheless a complete vestibular function tests and some audiological tests like PT audiometry and BERA are mandatory not so much for confirmatively diagnosing Vestibular Paroxysmia but more for ruling out other possible conditions.

High resolution MRI with 3D CISS (constructive interference in steady state) sequences of the brainstem with special instructions to the radiologist to look for any neurovascular compression in the first 1.5cm of the 8th cranial nerve can confirm and localise the vascular loop or neurovascular contact which is characterised by absence of detectable CSF layer between nerve and surrounding vessel in the required region i.e. proximal to the transition zone. In difficult to diagnose clinical conditions like suspected Vestibular Paroxysmia the MRI is also indicated for excluding differential diagnosis. Hence the HR MRI of the brainstem with 3D CISS sequence is a must in all suspected cases of Vestibular Paroxysmia. However just the radiological finding of neurovascular compression of the 8th cranial nerve without typical Vestibular Paroxysmia clinical features is not diagnostic of Vestibular Paroxysmia as in many normal patients also there is radiological evidence of neurovascular compression or the presence of a vascular loop in very close proximity of the 8th cranial nerve. Hence an incidental radiological finding of neurovascular compression involving the 8th cranial nerve is clinically insignificant and does not suggest vestibular paroxysmia. Not uncommonly in many patients the radiological evidence of neurovascular compression is present bilaterally and this is found not only in patients with typical VP symptoms but in many normal too. Only if both clinical features as well as radiological features are there only then the diagnosis of Vestibular paroxysmia is tenable.

Treatment:

No specific line of management has been proposed till date to treat VP. A therapeutic trial of low dose of carbamazepine (200-600 mg/day) or oxcarbazepine (300-900 mg/day) is recommended in all suspected cases taking care of the adverse effects that can be caused by these drugs. The patient should be strictly instructed to stop the drug and to seek urgent medical opinion if there is any untoward adverse effect as both Carbamazepine and to a lesser extent oxcarbazepine may sometimes induce life threatening allergic reactions. The response to these drugs during the attacks is a diagnostic test too and if there is gross improvement in the condition, then it more or less confirms the diagnosis of Vestibular Paroxysmia. Yet how long these drugs should be continued, is not well documented till date. The dose has to be titrated against the symptoms. Other drugs (phenytoin, lamotrigine, baclofen, topiramate, valproate, gabapentin) can be used if intolerance occurs to Carbamazepine or oxcarbazepine but no documented evidence is present till today about efficacy of those drugs in VP and their correct mechanism of action.

In medically intractable cases, surgery can be the option but should be avoided due to risk of dreaded complications like brainstem infarction (due to intra-op or post-op vasospasm). Also it is difficult to detect the side affected.

F. Tinnitus practice guidelines

Common Definitions: As per the Clinical Practice Guideline: Tinnitus by American Academy of Otolaryngology – Head and Neck Surgery

Tinnitus: The perception of sound when there is no external source of the sound

Primary tinnitus: Tinnitus that is idiopathic and may or may not be associated with sensorineural hearing loss

Secondary tinnitus: Tinnitus that is associated with a specific underlying cause (other than sensorineural hearing loss) or an identifiable organic condition

Recent onset tinnitus: Less than 6 months in duration (as reported by the patient)

Persistent tinnitus: 6 months or longer in duration

Bothersome tinnitus: Distressed patient, affected quality of life and/or functional health status; patient is seeking active therapy and management strategies to alleviate tinnitus

Nonbothersome tinnitus: Tinnitus that does not have a significant effect on a patient's quality of life but may result in curiosity of the cause or concern about the natural history and how it might progress or change

Evaluation of the patient

A. History to be taken covering the following issues–

a. Unilateral tinnitus

- i. Rule out focal auditory lesions like vestibular schwannoma or vascular tumours

b. Pulsatile tinnitus

- i. Rule out vascular tumor or systemic cardiovascular illness

c. Associated with Hearing Loss

- i. Very frequent association - find duration; type; severity; symmetry
 1. SNHL or CHL- frequently tinnitus is associated with SNHL
 2. Sudden SNHL – prompt treatment reqd
 3. Asymmetric – may be a Vestibular Schwannoma

d. New onset tinnitus – may diminish or disappear

e. Association with vertigo

- i. Meniere's disease
- ii. Superior Semicircular Canal Dehiscence
- iii. Vestibular Schwannoma
- iv. Other conditions

f. Noise exposure

g. Medications and ototoxic drugs exposure

h. Symptoms of Anxiety or Depression

- i. very important to assess the severity as it will guide the treatment and need for referral

i. Cognitive impairment

- i. Will affect the various tests

B. Physical Examination-

a. Objective tinnitus:

- i. Rarely, tinnitus can be heard by the clinician as well as the patient.
- ii. Vascular abnormalities and myoclonus.

b. Heart murmurs, carotid bruits, or vascular sounds

- i. Cardiovascular disease and vascular lesions may cause tinnitus.
- ii. Treatment of the underlying disease may help tinnitus symptoms
- iii. Cardiovascular disease requires appropriate evaluation and treatment.

- c. Focal neurologic signs -
 - i. may require referral or imaging
- d. Otorrhea
 - i. Sign of middle ear infection or otitis externa
 - ii. Treatment may improve tinnitus as well as associated hearing difficulties.
- e. Signs of other external or middle ear disease on examination and/or otoscopy
 - i. Simple problems such as cerumen impaction or otitis media can be detected. Cholesteatoma, glomus tumors, and other uncommon middle ear disorders can be detected by otoscopy.
- f. Head and neck masses
 - i. Prompt investigation required

C. Audiologic Evaluation

- 1) PTA- Properly masked AC & BC thresholds
- 2) SRT- agreement between PTA & SRT to check reliability
- 3) SDS
- 4) Tympanometry – if indicated
- 5) Tinnitus matching for frequency and intensity, maskability/ suppressability by external noise etc

D. Differentiate Bothersome Tinnitus from Nonbothersome

- a. Tinnitus Questionnaire and Tinnitus Effects Questionnaire (Hallam et al, 1988)
- b. Tinnitus Handicap Questionnaire (Kuk et al, 1990)
- c. Tinnitus Reaction Questionnaire (Wilson et al, 1991)
- d. Tinnitus Handicap Inventory (Newman et al, 1996)

E. Distinguish between patients with bothersome tinnitus of recent onset from those with persistent symptoms

F. Education & Counselling

- a. No Cure available
- b. Explain about tinnitus – what is it?
- c. Current hypothesis on pathophysiology
- d. Need to investigate and
- e. reassurance that it is not sinister if the requisite investigations have suggested so

G. Hearing Aid evaluation & recommendation if required

H. Sound Therapy

- a. Environment Enrichment devices
 - i. Table top sound devices
- b. Hearing Aids
 - i. Can help by partial masking effect
- c. Sound Generators
 - i. Musical pillows
- d. Combination Devices
 - i. Hearing Aid with a tinnitus masker

I. Cognitive Behavioural Therapy

The following are NOT recommended as they have not been found to be effective and is not evidence based

- A. Imaging studies are not to be recommended unless
 - a. Pulsatile tinnitus
 - b. Focal Neurological Abnormalities
 - c. Asymmetric Hearing loss
 - d. Head and neck swelling
- B. Medical Therapy of the following categories are not to be recommended as they have not been found to reduce the tinnitus
 - a. Anti depressants
 - b. Anxiolytics
 - c. Intratympanic Therapy

Should not be routinely recommended just for the tinnitus unless there is some separate additional indication which must be specified

C. Dietary Supplements

- a. Ginkgo
- b. Zinc
- c. Multivitamins
- d. Antioxidants

Should not be given just for tinnitus as they have not been found to be helpful

D. Acupuncture

E. Trans Cranial Magnetic Stimulation

5) NOMENCLATURE OF SYMPTOMS TO BE USED IN BALANCE DISORDERS

With some changes customized according to the terms that are used in medical communication in India and the neighboring countries, we will follow more or less the nomenclature proposed by the Barany Society. The clinician is to note that this nomenclature is related to reporting of symptoms only and this classification / nomenclature is in no way associated to the underlying causative pathology. The same pathology may sometimes present with any one or more of the symptoms enumerated below. Internationally the Classification of Vestibular Disorders is coded as ICVD-1 and reporting of the symptoms of vestibular disorders as reported by the patient is best done on the basis of this ICVD-1 system introduced by the Barany society as it maintains a uniformity of nomenclature. As per the nomenclature proposed by the Barany Society (Ref A. Bisdorff et al. / Classification of vestibular symptoms: Towards an international classification of vestibular disorders Journal of Vestibular Research 19 (2009) 1–13.) neurotological symptoms of balance disorder (with some changes) are broadly classified into the following categories viz:-

- 1) VERTIGO
- 2) DIZZINESS
- 3) UNSTEADINESS

- 1) VERTIGO:- The term vertigo encompasses those sensations where there is a feeling of movement/ motion either of the self *or of the surroundings**. As per the ICVD-1 classification movement of the visual surroundings without any self motion is termed as vestibulo-visual symptom or 'external vertigo' and the term 'vertigo' indicates only the feeling of self motion also called 'internal vertigo'. But this makes it unnecessarily too very complicated for use outside the vestibular community and vertigo is best used to denote both self motion as well as movement of the visual surroundings as explained in the next paragraph. Vertigo is the sensation of self-motion when no self-motion is occurring or the sensation of distorted or inappropriate self-motion during an otherwise normal head movement. If there is an actual movement / motion and the subject gets the appropriate sensation of the motion then it is not vertigo. The sensation of **motion** which is not rotatory / spinning in nature is termed as 'non-spinning vertigo' and the sensation of **spinning** which is a rotatory sensation is termed as 'spinning vertigo'. The term includes false spinning sensations (spinning vertigo) and also other false sensations of motion like swaying, tilting, bobbing, bouncing, or sliding (non-spinning vertigo). Terms like subjective vertigo, objective vertigo, true vertigo, false vertigo, rotatory vertigo etc. though often used in common medical parlance are not recognized clinical entities as per the Barany Society Classification of Vestibular symptoms (ICVD-1) referenced above and it is prudent for us to also follow so. These terms are hence best discontinued. Vertigo according to this classification of vestibular symptoms may be *spontaneous vertigo* when there is no known or obvious trigger (precipitating factor) or it may be *triggered vertigo* where there is a very definite relationship between some known stimulus and the vertigo. Examples of 'triggered vertigo' are e.g., *Positional vertigo* where change of head position in a particular direction causes the vertigo, *Head-motion vertigo* where there is a spinning sensation that is time-locked with head movement and occurs only when the head is moving and there is a perverted or distorted sense of self motion during actual self-motion, *Visually-induced vertigo* is a sense of self motion triggered by a movement of the visual field or a visual stimulus like seeing a moving train in a railway station, *Sound-induced vertigo* is vertigo triggered by an auditory stimulation which was previously termed as tulio-phenomenon, *Valsalva-induced vertigo* is vertigo triggered by increase of intracranial or middle ear pressure. Oscillopsia which is a to and fro up-down and even front back repetitive movement of the visual surroundings often described by patients as a jumping or bouncing of visual images while walking is actually a 'non-spinning vertigo'.

*As mentioned above another group of vertiginous sensations which have been grouped separately as vestibulo-visual symptoms and not included in VERTIGO in the ICVD-1 Barany society classification, consists of motion of the visual surroundings termed as 'external vertigo' or tilting of the visual images or a blurring of vision on head movements best included in this group of VERTIGO for convenience. The term external vertigo strictly indicates a sensation continuous movement of the visual surroundings usually jerky when there is actually no motion. This is the sensation of the movement of the visual surroundings only without any sense of self motion but very often but not always both co-exist. Conditions like *oscillopsia* detailed above, *visual lag* where there is a feeling that the visual surroundings move slightly (a second or so) later after a head movement and does not occur with the head movement which is normally expected and *visual blurring* immediately after or even during head movement fall under this group.

- 2) DIZZINESS is different from vertigo as per the Barany classification and Vertigo and Dizziness must be dealt with as separate symptoms. Dizziness is the jeopardised or perverted/ disturbed sense of spatial orientation without any sense of motion / spinning. There is neither any spinning nor any sense of motion in dizziness. Loss of spatial orientation is the inability of the subject to relate or to orient the self with the physical surroundings i.e., the three dimensional space surrounding the subject without any false sense of motion / spinning. A dizzy patient has problems in correctly determining his / her position in space and does not have any track of a physical reference point in relation to which the subject can determine his/ her position. Not uncommonly the patient can have both vertigo and dizziness together but these are two completely different entities. The same conditions that induce vertigo (all the different varieties like spontaneous / triggered and all the different triggers like position changing, loud sound etc.) can also induce dizziness and the description of dizziness is to be done in the same way as that of vertigo e.g., loud sound induced dizziness. Spatial disorientation as occurs in dizziness is different and should be distinguished from sensations like (1) impending loss of consciousness/ pre-faint, (2) mental confusion when the patient becomes mentally blank for a while or also (3) the sensation of depersonalisation when a subject feels detached from reality. None of these three symptoms are related to the vestibular system though quite often such patients report these symptoms as vertigo/ dizziness to the physician.
- 3) UNSTEADINESS: - is the term used to denote the feeling of instability or imbalance where the patient is helped when he /she gets a physical support. The patient gets a feeling of falling and /or may even fall but there is no sense of motion or that of spinning or of any spatial disorientation. The patient can have the feeling not only when walking but also when standing/ sitting. A patient of vertigo or of dizziness is not helped by getting some physical support like holding to something but if a patient has unsteadiness the symptoms disappear completely by holding on to something. Though vertigo and dizziness are primarily vestibular symptoms unsteadiness may be present in persons with normal vestibular function and may be caused by non-vestibular conditions like Like dizziness, unsteadiness may coexist with the other symptoms. Unsteadiness has been sub-classified in the ICVD-1 as:-
 - 1) Directional pulsion i.e., being pushed to one particular side. If the feeling is that of being pushed to the left or right it is termed as latero-pulsion to left / right. If the directional pulsion is backwards it is called retro-pulsion and if to the front it is called antero-pulsion
 - 2) Near falls are the feeling of an impending fall but which has been prevented or arrested by catching onto something and a complete fall has not occurred. But unsteadiness may be due to non-vestibular causes also. Only the balance system related near falls are in this category.
 - 3) Complete falls are when the fall has not been prevented by catching on to something.

All balance disorder symptoms are best reported under the headings of vertigo/ dizziness / unsteadiness and if possible then further sub-classified as e.g., spontaneous internal vertigo or triggered positional vertigo, unsteadiness with retropulsion , loud sound triggered dizziness etc.