6. MATERIAL AND METHOD
6. MATERIAL AND METHODS

'Computer-Assisted Diagnosis in NeurOtology' (CADINO), an experimental computer program, which is capable of making diagnoses in cases of dizziness, includes various causes of dizziness (Table 6.1 and 6.2). It was developed and evaluated during the period extending from January 2006 to February 2008 in the Department of E.N.T., B.J. Medical College Ahmedabad. It was developed in Microsoft ® Word 2000 using hyperlinks. Files can be saved and opened in any web-designing tool. Prospective study of the diagnostic accuracy of CADINO consultations was done in 35 patients, 8 simulated cases and 7 case reports from journals. The study included even the opinions of the ENT surgeons before and after consultation. Patients were from the Department of E.N.T., B.J. Medical College Ahmedabad. Simulated cases were presented in the monthly academic meeting by the members of Anand-Kheda chapter of Gujarat State Branch (GSB) of Association of Otolaryngologists of India (AOI). Case reports were taken randomly from the Vertigo Viewpoint journal. Thirty-five patients of dizziness, 11 ENT residents and 14 ENT surgeons (8 teachers and 6 senior private consultants) participated in the evaluation of CADINO.

The components of CADINO are Knowledge base, Inference engine, and User interface. The knowledge base is a collection of the diseases, which can lead to dizziness and are needed to solve problems in the patients of dizziness. The inference mechanism is a computer program, that given a patient details, uses the information in the knowledge base to generate list of differential diagnosis about the dizziness. While these two components are viewed here as being conceptually distinct, they are interwoven in the system. The CADINO knowledge base was designed to permit the program to construct and resolve differential diagnoses.

KNOWLEDGE BASE

The knowledge base incorporates individual disease profiles, which consists of historical items, symptoms, physical signs, laboratory abnormalities and treatment. An additional concern is that certain diseases may present differently in some patients. For example,
<table>
<thead>
<tr>
<th>Central Vestibular Disorders</th>
<th>Spino cerebellar Ataxias</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vascular</td>
<td></td>
</tr>
<tr>
<td>Ischemic Stroke / Transient Ischemic Attack</td>
<td></td>
</tr>
<tr>
<td>Dissection of Vertebral Artery</td>
<td></td>
</tr>
<tr>
<td>Stenosis of Proximal Subclavian Artery – Subclavian Steal Syndrome</td>
<td></td>
</tr>
<tr>
<td>Verteobasilar Transient Ischemic Attacks</td>
<td></td>
</tr>
<tr>
<td>Lateral Medullary Infarction – Wallenberg’s Syndrome (Occlusion of vertebral artery or posteroinferior cerebellar artery)</td>
<td></td>
</tr>
<tr>
<td>Lateral Pontomedullary Infarction</td>
<td></td>
</tr>
<tr>
<td>Vestibular-masseter Syndrome</td>
<td></td>
</tr>
<tr>
<td>Cerebellar Infarction</td>
<td></td>
</tr>
<tr>
<td>Hemorrhage: Brainstem, Cerebellar, Parietoinsular Cortex/ Superior or medial Temporal Gyri</td>
<td></td>
</tr>
<tr>
<td>Migraine</td>
<td></td>
</tr>
<tr>
<td>Basilar Migraine &amp; Vestibular Migraine</td>
<td></td>
</tr>
<tr>
<td>Episodic Ataxias: Type 1 &amp; 2 (EA-1 &amp; 2)</td>
<td></td>
</tr>
<tr>
<td>Benign Paroxysmal Vertigo of Childhood</td>
<td></td>
</tr>
<tr>
<td>Benign Recurrent Vertigo</td>
<td></td>
</tr>
<tr>
<td>Benign Paroxysmal Torticollis of Infancy</td>
<td></td>
</tr>
<tr>
<td>Frontal Gait Disorder of Elderly</td>
<td></td>
</tr>
<tr>
<td>(Gait Apraxia or Lower-half Parkinsonism – Multiple Lacunar Infarcts)</td>
<td></td>
</tr>
<tr>
<td>Superficial Siderosis</td>
<td></td>
</tr>
<tr>
<td>Inflammatory</td>
<td></td>
</tr>
<tr>
<td>Multiple Sclerosis (MS)</td>
<td></td>
</tr>
<tr>
<td>Cerebellitis</td>
<td></td>
</tr>
<tr>
<td>Susac Syndrome</td>
<td></td>
</tr>
<tr>
<td>Infectious</td>
<td></td>
</tr>
<tr>
<td>Intracranial Complications of Otitis Media</td>
<td></td>
</tr>
<tr>
<td>Meningitis</td>
<td></td>
</tr>
<tr>
<td>Cerebellar Abscess (Fungal or Bacterial)</td>
<td></td>
</tr>
<tr>
<td>Lyme Disease</td>
<td></td>
</tr>
<tr>
<td>Neoplastic</td>
<td></td>
</tr>
<tr>
<td>Brainstem and cerebellum</td>
<td></td>
</tr>
<tr>
<td>Paraneoplastic syndromes</td>
<td></td>
</tr>
<tr>
<td>-Paraneoplastic Cerebellar Degeneration</td>
<td></td>
</tr>
<tr>
<td>-Paraneoplastic Opsoclonus - Myoclonus</td>
<td></td>
</tr>
<tr>
<td>Craniocervical Junction Disorders</td>
<td></td>
</tr>
<tr>
<td>Chiari Malformation</td>
<td></td>
</tr>
<tr>
<td>Inherited Ataxias</td>
<td></td>
</tr>
<tr>
<td>Autosomal Recessive: Friedreich Ataxia</td>
<td></td>
</tr>
<tr>
<td>Autosomal Dominant:</td>
<td></td>
</tr>
<tr>
<td>Metabolic</td>
<td></td>
</tr>
<tr>
<td>Wernicke’s Encephalopathy</td>
<td></td>
</tr>
<tr>
<td>Diabetes</td>
<td></td>
</tr>
<tr>
<td>Vitamin B-12 Deficiency</td>
<td></td>
</tr>
<tr>
<td>Vitamin E Malabsorption</td>
<td></td>
</tr>
<tr>
<td>Celiac Disease</td>
<td></td>
</tr>
<tr>
<td>Hypothyroidism</td>
<td></td>
</tr>
<tr>
<td>Hypoparathyroidism</td>
<td></td>
</tr>
<tr>
<td>Hypoglycemia</td>
<td></td>
</tr>
<tr>
<td>Hyperventilation</td>
<td></td>
</tr>
<tr>
<td>Toxic</td>
<td></td>
</tr>
<tr>
<td>Medications: Methotrexate</td>
<td></td>
</tr>
<tr>
<td>Alcohol</td>
<td></td>
</tr>
<tr>
<td>Degenerative</td>
<td></td>
</tr>
<tr>
<td>Parkinson’s Disease</td>
<td></td>
</tr>
<tr>
<td>Progressive Supranuclear Palsy</td>
<td></td>
</tr>
<tr>
<td>Multiple Systems Atrophy</td>
<td></td>
</tr>
<tr>
<td>Epilepsy</td>
<td></td>
</tr>
<tr>
<td>Partial Seizures</td>
<td></td>
</tr>
<tr>
<td>Trauma</td>
<td></td>
</tr>
<tr>
<td>Post-concussion Syndrome</td>
<td></td>
</tr>
<tr>
<td>Physiologic</td>
<td></td>
</tr>
<tr>
<td>Motion Sickness</td>
<td></td>
</tr>
<tr>
<td>Mal de Debarquement Syndrome</td>
<td></td>
</tr>
<tr>
<td>Psychophysiological</td>
<td></td>
</tr>
<tr>
<td>Chronic Anxiety</td>
<td></td>
</tr>
<tr>
<td>Panic Disorder</td>
<td></td>
</tr>
<tr>
<td>Phobic Postural Vertigo</td>
<td></td>
</tr>
<tr>
<td>Global Cerebral Hypoperfusion</td>
<td></td>
</tr>
<tr>
<td>Vasovagal Presyncope</td>
<td></td>
</tr>
<tr>
<td>Reduced Cardiac Output</td>
<td></td>
</tr>
<tr>
<td>Autonomic Insufficiency</td>
<td></td>
</tr>
<tr>
<td>Hypovolemia</td>
<td></td>
</tr>
<tr>
<td>Multisensory Disturbances</td>
<td></td>
</tr>
<tr>
<td>Peripheral Neuropathy</td>
<td></td>
</tr>
<tr>
<td>Cervical or Thoracic Myelopathy</td>
<td></td>
</tr>
<tr>
<td>Visual Loss</td>
<td></td>
</tr>
<tr>
<td>Aging: Cerebellar atrophy and diffuse small vessel ischemia</td>
<td></td>
</tr>
<tr>
<td>Ocular Motor Disorders</td>
<td></td>
</tr>
<tr>
<td>Superior Oblique Myokymia</td>
<td></td>
</tr>
<tr>
<td>Voluntary Nystagmus</td>
<td></td>
</tr>
</tbody>
</table>
Table 6.2. Peripheral Vestibular Disorders

- **Infectious**
  - **Viral**
    - Viral Labyrinthitis (Vestibular Neuritis)
    - Measles, Mumps, Infectious mononucleosis, Herpes zoster (VZV) Oticus (Ramsay Hunt Syndrome)
  - **Bacterial**
    - Toxic or Serous labyrinthitis and Suppurative labyrinthitis due to Acute otitis media, Chronic otitis media: Cholesteatoma (through labyrinthine fistula), Bacterial meningitis, Ear surgeries
  - Labyrinthine fistula due to Cholesteatoma
- **Fungal**
  - Systemic Otomycosis
    - Chronic infectious / granulomatous
      - Syphilis (Early, Late stage Acquired & Congenital)
      - Lyme disease
      - Tuberculosis
    - Granulomatous
      - Langerhans cell histiocytosis
      - Wegner's granulomatosis
      - Sarcoidosis
  - **Traumatic**
    - Nonpenetrating trauma
      - Labyrinthine Concussion: Blunt Head trauma, Inner ear barotrauma
      - Benign paroxysmal positional vertigo (BPPV)
    - Blast Trauma: An open handed slap to the ear, Explosions
  - Penetrating Trauma (Violation of otic capsule)
    - Temporal bone fracture: Transverse, Longitudinal (Labyrinthine concussion), Oblique / Mixed
    - Introduction of foreign bodies: Hair pin, Bullet
  - Ear Surgery – Stapedectomy
  - Barotrauma
    - Alternobaric trauma
    - Atmospheric inner ear barotraumas
    - Inner ear decompression sickness
    - Isobaric gas counterdiffusion sickness

- **Metabolic**
  - Hyperviscosity syndromes
  - Diabetes mellitus
  - Hyperlipoproteinemia
  - Hypothyroidism
  - Hormonal fluctuations: Premenstrual, Perimenopausal, Oral contraceptive, Estrogen replacement therapy
  - Otosclerosis

- **Collagen Vascular Disorders**
  - Autoimmune inner-ear disease
  - Primary
  - Secondary
  - Nonsyphilitic interstitial keratitis (Cogan’s syndrome)
- **Ischemic**
  - Small labyrinthine vessels – Isolated infarction of vestibular labyrinth
  - Occlusion of labyrinthine artery – Sudden hearing loss and vertigo
- **Drug-induced**
  - Aminoglycoside antibiotics
  - Antineoplastic agents: Cisplatin
- **Endolymphatic Hydrops**
  - Meniere’s disease (Drop attacks & Lermoyez attacks)
  - Delayed Endolymphatic Hydrops
  - Recurrent Vestibulopathy
- **Neoplastic**
  - Cerebellopontine angle tumors
    - Acoustic neuroma, Meningioma, Epidermoid, Leukemia
  - Temporal bone tumors
  - **Miscellaneous**
    - Ear wax
    - Familial Vestibulopathy
    - Recurrent Vestibulopathy
    - Vestibular neuritis
    - Sudden sensorineural hearing loss
    - Perilymphatic fistula (Inner ear fistula): Round or oval window fistula
    - Benign paroxysmal positional vertigo (BPPV)
    - Superior semicircular canal dehiscence syndrome
    - Congenital cholesteatoma
perilymphatic fistula may present as transient vertigo or imbalance as well as similar to Meniere’s disease.

Solution of classification problem entailed the development of algorithms that permit CADINO to construct problem areas, which are selected groups of observed findings. The CADINO knowledge base encompasses about 100 causes of dizziness (Table 6.1 and 6.2). The disease profiles have been derived from interaction with the ENT surgeons especially Dr. Vikas Sinha (Professor & Head, ENT Department, BJ Medical College, Ahmedabad) and authentic postgraduate level textbooks of otolaryngology especially Fourth edition (2005) Volume IV of Cummings Otolaryngology Head & Neck Surgery published by Elsevier Mosby Philadelphia USA.

In addition to disease profiles, the knowledge base details relations among diagnoses and among manifestations. The knowledge base contains links between the related diseases, which come under similar pathophysiological states. The links are used to express causality or a predisposition. CADINO formulates and resolve problem areas serially. The total number of links among the 100 diagnoses in knowledgebase is about 1000. The manifestations of diseases in CADINO knowledge base are not independent. The knowledge base has properties of each manifestation that specify how its presence or absence may influence the differential diagnoses. During the development of CADINO, many cases, both simple and complex, were presented to the system in order to evaluate and improve the knowledge base and the diagnostic computer program. The knowledge base of CADINO was accordingly altered after such consultations. These cases were not included in the evaluation study.

KNOWLEDGE REPRESENTATION

In developing CADINO, perhaps the most challenging part was to translate pathophysiological knowledge of dizziness into abstract representation that could be processed by computer. It involved many approaches.

I have employed two distinct methods of knowledge elicitation:

1. Classification or scaling method.
2. The traditional Interview based approach.

The CADINO consists of following two types of knowledge:
1. Declarative knowledge consists of factual knowledge of pathophysiology of dizziness.

2. Procedural knowledge is the skill of knowing how to use declarative knowledge.

Declarative knowledge is derived from interaction with the ENT surgeons especially Dr. Vikas Sinha and authentic postgraduate level textbooks of otolaryngology especially Cummings Otolaryngology Head & Neck Surgery (2005). Expressing medical problem-solving knowledge as a set of algorithm was a fairly difficult task, as the diagnostic knowledge is intuitively known to otoneurologist and found in medical books as descriptive data.

Problem-solving algorithms represent the intellectual core of CADINO. The behavior of CADINO results mainly from the formation of problem areas through a partitioning algorithm and the conclusion of diagnoses within problem areas, using strategy of diagnosis by exclusion. Decision Analysis is a formal discipline for making decisions that in many respects resembles the informal strategies of clinicians. In CADINO it is presented as a decision tree with several pathways along each of which are several nodes or decision points. Each intervention is a decision node. The tree is progressively built, from the findings of the dizziness patient. The tree is constructed in sufficient detail to make the representation realistic, and yet is constrained to prevent it from becoming unmanageably large.

CADINO attempts to form an appropriate differential diagnosis in cases of dizziness. We routinely construct differential diagnoses on the basis of etiology (such as infection, trauma, congenital, neoplasm, iatrogenic, idiopathic) or the site of lesion central and peripheral vestibular diseases (Table 6.1 and 6.2). A clinician thus narrows the set of possible diagnoses from all known diseases. The method of diagnosis by exclusion is employed to resolve each differential diagnosis. This strategy represents an attempt to model clinicians’ behavior.

CADINO basically follows a symptom-based classification, which is based on presumed pathophysiologic mechanisms, to evaluate the cases of dizziness that includes many different sensations such as vertigo (a sense of motion of person or surrounding), disequilibrium, and presyncope (near-faint, light-headedness). Some diseases have both central as well as peripheral vestibular features. The follow up CADINO consultations
may give a new list of differential diagnoses due to the additions of new findings during the temporal profile and natural history of CNS disorders.

A patient’s history often provides enough information to diagnose a cause for various types of dizziness. The CADINO program tries to get important history information that usually leads to identification of a category or process to which patient’s symptoms are attributable. CADINO considers the following key features of the history:

1. Description of the dizziness:
   a. Unsteadiness and Disequilibrium
   b. Lightheadedness
   c. Vertigo and oscillopsia
   d. Presyncope

2. Episodic or continuous

3. Duration of the individual attack: Seconds, Minutes, Hours, Days, or Months

4. Frequency: Daily vs. monthly.

5. Effect of head movements: worse, better, or no effect

6. Specific positions that induce vertigo e.g. rolling onto the side in bed.

7. Preceding history of
   a. Trauma: Physical, barotraumas, surgical
   b. Medicines: For hypertension, hyperglycemia and cardiac arrhythmias

8. Medical conditions: Hypothyroidism, diabetes mellitus, anemia, autoimmune diseases, and hypoperfusion of brain from postural hypotension or cardiac arrhythmia

9. Psychogenic disorders: Anxiety and panic disorders and agoraphobia

10. Triggering events, stimuli (sound or pressure), or movements

11. Associated / concomitant symptoms
    a. Ear: Discharge, pain, hearing loss, tinnitus, ear fullness
    b. Eye: Diplopia, vision loss
    c. Headache
    d. CNS: Paralysis and paresthesias, dysarthria, dysphagia
    e. Sweating, dyspnea and palpitation
HOW CADINO LOOKS?
CADINO employs an organized method for history taking and physical examination leading to the identification of specific abnormalities. Its approach to patients is built on an understanding of pathophysiology of dizziness. The CADINO program asks data on patient findings necessary for diagnostic work with dizziness patients. The following steps are taken during a CADINO diagnostic consultation. In order to improve the efficiency, questions in small groups are asked by the system. The level of questioning is escalated step by step. The program changes focus from one problem area to another as per the selection of the option. This method of constructing differential diagnoses gives CADINO seemingly intelligent behavior.

CADINO assumes that only one disease process is present at a time and user can choose only one option from many given choices. But if clinician feels that patient has multiple problems at a given time and more than one options are correct, than user will have to run the program again and select the options accordingly. CADINO can play role of an electronic textbook as well as diagnostic consultant.

When the program is started, options are displayed on the screen. One most relevant finding of the patient can be chosen directly from the menu. An interactive dialogue is then started that will lead to a possible diagnosis based on the findings of the dizzy patient. The line of inquiry is based on the users identification of one most relevant finding of the patients. The user cannot choose more than one finding. It is best to take time and choose the one most pertinent finding. Some findings have additional description, which follow that finding. Hearing loss can be conductive or sensorineural, preexisting or recent, sudden or gradual onset, and progress rapidly, gradually or fluctuating. Some choices off the category menus are technically not 'true symptoms.' These include important personal risk factors such as Diabetes, Hyperlipidemia, Hypertension or Smoking history.

EVALUATION OF CADINO
The CADINO was evaluated to highlight its strengths and weaknesses. Its clinical acumen was compared with that of residents, E.N.T. consultants and teachers. The study included even the opinions of the users before and after consultation. The evaluation was not intended to validate CADINO for the clinical use.
Thirty-five patients of dizziness, 11 ENT residents and 14 ENT surgeons (8 teachers and 6 senior private consultants) participated in the evaluation of CADINO. Prospective study of the diagnostic accuracy of CADINO consultation was done in 35 patients, 8 simulated cases and 7 case reports from journals. Educational consultations,
which were done for seeing pertinent disease profile, causes of key findings, or relationships between findings, were not included in the analysis.

Consultations were solicited from otolaryngology department of BJ Medical College Ahmedabad. The patients in whom no cause of dizziness could be ascertained (such as vestibulopathies of unknown etiologies and presumed malingering) and who have multiple causes of dizziness (such as elderly patients) were not included in the study. The chief medical resident identified the cases of dizziness or the attending faculty member or house staff directly asked for CADINO consultation. The consultations were carried out by residents and faculty members those were conversant with the use of CADINO. The system used to prompt users for additional information. The faculties reviewed the patients record and selected the patient’s findings from the CADINO program. The system then generated a list of diagnoses along with the suggestions for additional history, physical or laboratory items.

Any additional information obtained through pertinent history, directed physical examination and investigations helped in arriving the final diagnosis. CADINO’s suggestions were noted regarding any potentially discriminating items of information not yet obtained. Residents were free to act on the CADINO’s suggestions as per the directions of their faculties.

Before the consultation, the concerned resident was requested to fill out a brief preconsultation form summarizing the available findings and recording their differential diagnoses. Faculties were also requested to note their diagnoses before CADINO consultation. (Box 6.1)

All patients who underwent CADINO consultation from September 2007 to February 2008 were included in the study. The protocol followed for the patients of CADINO consultation was a detail history and thorough examination. Provocation tests such as Hallpike and hyperventilation tests were done if indicated. Laboratory investigations, audiological testing, and imaging were done if only strongly indicated.

Users were asked to record the following items after CADINO consultation:

- CADINO’s differential diagnoses
- Any change in the management and investigation plan
- Rating of the educational value (Helpful, neutral, not helpful)
- Rating of the value in patient management (Helpful, neutral, not helpful)
The patients were followed until a diagnosis was definitively established through history and examination. Laboratory or imaging studies were done if strongly indicated. The diagnosis was finally established with the consultation of professor and head department of ENT. If sufficiently convinced of the diagnosis specific treatment would be started. The diagnostic accuracy of the residents, faculties and program and the impact of CADINO consultation were assessed comparing the final established diagnosis.

Evaluation study with 8 simulated cases was done in one monthly academic meeting by the members of Anand-Kheda chapter of Gujarat State Branch (GSB) of Association of Otolaryngologists of India (AOI). The simulated cases were presented by the individual members, who filled up the preliminary form (Box 6.2) and approved by the rest of members present in the meeting. Diagnostic decisions made by CADINO were classified as correct that was agreed by rest of the members. After CADINO consultation, present members were requested to record the rating of the educational value (Helpful, neutral, not helpful) and patient management (Helpful, neutral, not helpful).

<table>
<thead>
<tr>
<th>Box 6.2. Form for Simulated Cases to be filled by the member of AOI</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Computer-Aided Diagnosis In Neurotology (CADINO)</strong></td>
</tr>
<tr>
<td><strong>Clinical Evaluation: Usefulness, Functionality and Effectiveness</strong></td>
</tr>
</tbody>
</table>

Doctor:

Diagnosis of Simulated Case:

CADINO Diagnosis:

Consensus Diagnosis:

Educational value of CADINO: Helpful / Neutral / Not helpful

CADINO for Patient’s Management: Helpful / Neutral / Not helpful

Time taken for CADINO Consultation (in minutes)

Doctor’s Comments / Suggestions regarding CADINO:

The clinical acumen of CADINO was also evaluated in 7 case reports, which were taken randomly from the Vertigo Viewpoint journal that is published by Excerpta Medica.
Asia Ltd. Hong Kong. The journal is supplied, complimentary to doctors by Janssen-Cilag/Johnson & Johnson Ltd. During the trial, only the published findings were used and knowledge base of CADINO was not altered.
Appendix: Clinical Features of Diseases

One must determine whether the patient has a peripheral or central cause of vertigo. The typical clinical presentations of the various causes of dizziness, which aid in making this distinction, are described here under two heads peripheral and central vestibular disorders. The disease profiles and declarative knowledge have been derived from interaction with the ENT surgeons especially Dr. Vikas Sinha (Professor & Head, ENT Department, BJ Medical College, Ahmedabad) and authentic peer reviewed journals and postgraduate level textbooks of otolaryngology especially Fourth edition (2005) Volume IV of Cummings Otolaryngology Head & Neck Surgery published by Elsevier Mosby Philadelphia USA. Problem-solving algorithms, which represent the intellectual core of CADINO, were derived from the declarative knowledge.

PERIPHERAL VESTIBULAR DISORDERS

1. INFECTIOUS
   a. Viral
      The widespread use of vaccines against rubeola (measles), rubella, mumps, and varicella (chickenpox) will dramatically decrease the cases of vertigo and hearing loss, which occur due to these viruses.

      i. Viral Labyrinthitis or Vestibular Neuritis\textsuperscript{121, 303, 306, 312} (Syn. Vestibular Neuritis or Neuronitis – Acute Viral Labyrinthitis)
         This is the second most common cause of peripheral vestibular vertigo
         \textit{Etiology}
         50% patients have preceding H/O URI and sinusitis. Following microorganisms, which cause vertigo, have been suggested but not confirmed: mumps virus, \textit{Varicella zoster} virus (VZV), influenza, \textit{Epstein bar} virus (EBV) of infectious mononucleosis, herpes simplex virus (HSV), \textit{Borrelia burgdorferi} (Lyme disease) and \textit{T. pallidum}.
         \textit{Clinical}\textsuperscript{243}
         • Often patients have dramatic sudden onset of vertigo and attendant vegetative symptoms (such as nausea, vomiting and sweating) with gradual, definite improvement throughout the course.
• Occasionally patients have bouts of attacks over several weeks. Few patients complain of stuffiness in their ear.
• Horizontal or horizontal-rotary nystagmus is towards uninvolved side.
• Imbalance caused by rapid head movement may lasts for months after the resolution of acute disease.
• Paroxysmal positional vertigo occurs subsequently in some patients. Some patients have recurrent attacks (transient positional vertigo triggered by sudden head movements) for years, which are usually less intense.
• Bilateral disease though rare must be considered in DD of bilateral vestibular loss.

Investigations
• Caloric responses are decreased or absent and identify the side of involvement.
• CSF and EEG: Normal
• Many patients have preceding infection of Herpesvirus (increased serum titer).
• Though there is no subjective hearing loss audiograms usually show hearing loss in high frequencies (10-15 kHz).

DD
• Hemorrhage parietoinsular cortex or superior or medial temporal gyri (vestibular cortex)
• Multiple Sclerosis
• Cerebellar lesions – abscess, hemorrhage, infarction
• Vascular lesions of labyrinth.

Prognosis
Vertigo usually lasts many hours to 1 to 2 weeks and then spontaneously disappears over weeks to months. Prolong antivertiginous therapy delays the recovery time.

ii. Measles Labyrinthitis
Rubeola virus causes 3% to 10% of all acquired deafness in unvaccinated children.

Clinical
– Abrupt onset of hearing loss (usually permanent, bilateral asymmetric high frequency and varying from mild-moderate to severe) at the time of rash.
– May be accompanied by vertigo and tinnitus.
– Diminished or absent caloric response.

Diagnosis
– Rubeola virus from throat cultures
– Rubeola virus RNA
– Rubeola virus antigen by immunofluorescent staining of exfoliated epithelial cells from oral cavity, pharynx, and conjunctiva.
– Fourfold or more rise in serum measles antibodies titer between acute and convalescent phase.
iii. Mumps Labyrinthitis
Mumps vaccine has reduced the incidence.\textsuperscript{108}

\textit{Clinical}\textsuperscript{257}
- Rapid and unilateral high frequency hearing loss (often profound and permanent) usually develops toward the end of parotitis but can occur from a subclinical infection without parotid swelling.
- Tinnitus and fullness are common.
- Some patients develop vertigo, which usually resolves over several weeks with permanent diminished or absent caloric response.

\textit{Diagnosis}
- Isolation of mumps virus or its RNA from throat or CSF.
- Fourfold or more increase in serum mumps antibodies between acute and convalescent phase.

iv. Herpes zoster (Varicella-Zoster Virus) – Ramsay Hunt Syndrome
Herpes Zoster Oticus is caused by Varicella-Zoster Virus (VZV) that remains latent in geniculate ganglia for decades after acute chickenpox. VZV starts replicating first in geniculate ganglia and then travels down and affect facial nerve, inner ear, spiral and vestibular ganglia.

\textit{Clinical}\textsuperscript{147, 322}
- Painful vesicles appear on and behind pinna and in external auditory canal.
- Unilateral facial palsy and deep ear pain manifest 1-2 days later. Facial paresis usually recovers over weeks.
- About 25% patients have vertigo, nystagmus, tinnitus, and hearing loss.
- Sensorineural hearing loss (SNHL) occurs only in 6% of cases.
- Caloric responses are either decreased or absent.

\textit{Diagnosis}
- Clinical
- Histopathological examination (HPE): Multinucleated giant cells in vesicle scrapings.
- VZV from vesicle fluid
- VZV DNA by PCR assay in vesicle fluid or CSF

b. Bacterial and Fungal\textsuperscript{242}
i. Toxic Labyrinthitis\textsuperscript{263}

\textit{Causes}
- Systemic febrile illnesses such as pneumonia or influenza.
- Fatigue and allergy of internal ear.
- (Viral and ototoxic drugs including quinine, lead, arsenic, zinc)

\textit{Clinical}
- Vertigo develops gradually and reaches maximum severity within 48 hours and then gradually subsides over several weeks. Nausea
and vomiting may be present. Head movement may worsen the vertigo.

- Hearing loss and tinnitus are absent.
- After months or years hearing loss and tinnitus may appear, leading to a definite diagnosis of Meniere’s disease.

ii. **Serous Labyrinthitis**

Transient nonpurulent inflammation or chemical irritation of the inner ear, which usually does not result in any permanent damage. If diagnosed and treated successfully before the development of suppurative labyrinthitis, the prognosis is excellent. Hearing and vestibular dysfunction are entirely reversible. The perilymph is involved but not the endolymph.

**Causes**

- Causes of Circumscribed labyrinthitis (labyrinthine fistula)
- Bacterial or viral toxin invasion through round or oval window: All types of otitis media and mastoiditis.
- Blood-borne infection
- Meningeal inflammation
- Ear surgery: Fenestration and stapedectomy

**Clinical**

- Spontaneous vertigo in cases of middle ear infection usually indicates serous labyrinthitis. Vertigo is more severe than labyrinthine fistula but less severe than suppurative labyrinthitis.
- Irritative type of nystagmus (Hyperactive labyrinthitis): Quick component is directed toward the affected ear.
- If the condition is secondary to labyrinthine fistula (LF), fistula sign will be positive. Though there occur worsening of LF symptoms. Vertigo is less severe because some compensation develops during the inflammation of LF.
- Hearing may be impaired but not markedly. If useful hearing is present, then suppurative labyrinthitis has not developed. Insidious high-tone SNHL is a frequent accompaniment of CSOM even without vertigo.
- Caloric tests usually reveal diminished vestibular response.

iii. **Suppurative (Purulent) Labyrinthitis**

- Routes of infection: Tympanogenic (otitis media, mastoiditis, petrositis, temporal bone #, penetrating injury or surgery through tympanic membrane, tumor), meningeogenic and hematogenic (from distant or systemic infection or from adjacent areas such as meningitis, encephalitis or brain abscess). Usually due to Acute otitis media, cholesteatoma and meningitis.
- Complete hearing loss (permanent) and acute vertigo, which slowly resolves over weeks to months. H/O Ear discharge / ear pain / preceding cold and cough.

**Predisposing factors**

- Mondini deformity
- Enlarged vestibular aqueduct
• Stapes surgery
• Weak or dehiscent internal auditory canal (IAC) opening or foramina into the medial aspect of labyrinth.
• Patent cochlear aqueduct
• Labyrinthine fistula

3 Types or Stages
• I - Acute (manifest) stage (1-2 weeks):
  o Vestibular symptoms peaking during the first few days gradually subsides after the first week. Patient lies quietly on the affected ear and cannot stand or sit. Even slight head movements produce vomiting.
  o Complete hearing loss, past pointing and fall occur toward the diseased side.
  o Headache, pain and fever are absent.
  o Horizontal rotary nystagmus has the quick component on affected side for the first day (irritative) but than toward the opposite side (paralytic).
  o Unilateral labyrinthitis produces more severe vestibular upset than bilateral meningogenic labyrinthitis.

DD:
Cerebellar abscess, vascular occlusion, and multiple sclerosis.

• II - Chronic (latent or fibrous) stage (1-6 weeks):
  Milder vestibular upset and positional vertigo.
  Absence of both cochlear and vestibular function.
  Bilateral lesions lead to difficulty in walking especially in the dark or on a soft surface.

• III - Healed (compensated or ossification) stage (Labyrinthitis ossificans): New bone forms over several months. Many years are required for the development of complete labyrinthitis ossificans.

  1. Tymanogenic Bacterial Labyrinthitis:
     Acute otitis media, Chronic otitis media: Cholesteatoma (through labyrinthine fistula) and Otomycosis

Clinical
  o Rapid onset (½ to 1 hour) of tinnitus, whirling vertigo, pallor, diaphoresis (sweating), nausea and vomiting.
  o Symptoms remain severe and may not respond to any treatment for 8 to 12 hours.
  o Severity improves during next few days but head motion evokes severe vertigo and nausea.
  o CNS compensation occurs over next 2 or 3 weeks.
  o Brisk jerky nystagmus directed toward the normal ear.

DD
Middle ear effusion is the most common cause of vestibular disturbance in children. Glue ear children may walk clumsily or trip easily and this dizziness clear with myringotomy and grommets. The dizziness may develop from transient negative middle ear pressure changes associated with displacement of
round window membrane, leading to secondary movement of perilymph.

Serous labyrinthitis can occur secondary to the superadded infection of middle ear effusion.

2. Meningogenic Bacterial Labyrinthitis:

Meningitis

Bacteria, fungi or inflammatory cells reach the basal turn of scala tympani from the subarachnoid space by way of a patent cochlear aqueduct (often patent in small children and closes as the skull grows) or up the internal auditory canal reaching the cochlear modiolus passing through perineural and perivascular spaces.

About 33% of all acquired SNHL are caused by meningitis. Hearing loss varies by causative organisms: *Streptococcus pneumoniae*, 20%; *Haemophilus influenzae*, 12%; and *Neisseria meningitides* 5%.

*Clinical*

- Vertigo, nausea, vomiting, and ataxia often develop regardless of hearing loss.
- Deafness (usually profound and permanent involving all frequencies), which may be either unilateral or bilateral, usually occurs early.

iv. Labyrinthine Fistula (Circumscribed Labyrinthitis, Paralabyrinthitis, and Perilabyrinthitis)

Loss of endochondral bone without loss of perilymph occurs usually due to Cholesteatoma (mainly horizontal semicircular canal). Inflammation of a discrete portion of bony labyrinth and endosteum without the involvement of membranous labyrinth. Labyrinthine fistula is usually accompanied by erosion of bony fallopian canal and tegmen. So surgeon should be careful.

Patient may remain asymptomatic.

*Causes*

- Cholesteatoma – Most common
- Chronic Otitis Media
- Congenital Syphilis
- Carcinoma
- Glomus Jugulare Tumors
- Fenestration operation for otosclerosis – No inflammation

*Clinical*

- Secondary to cholesteatoma
- Brief periods of imbalance, disequilibrium, or vertigo but normal equilibrium most of the time.
- Momentary imbalance on pushing external ear canal with washing cloth (Fistula sign). Transient vertigo induced by washing the ear or by pushing on the tragus.
- Tullio’s phenomenon; loud sounds provoke brief imbalance.
• Fistula test positive: Positive pressure causes utriculopetal movement of endolymph in lateral SCC, which leads to conjugate deviation of eyes away from the disease side, while vice versa occurs when negative pressure is created in EAC.
• CT Scanning in bone algorithm: Bone erosion of lateral SCC.

c. **Immunocompromised Patients**

Systemic invasive mycoses (Aspergillosis, Mucormycosis, Candidiasis, Cryptococcosis, Coccidioidomycosis, Histoplasmosis) involve temporal bone in cases of defective host defenses (diabetic ketoacidosis, chemotherapy, corticosteroids, AIDS). Chronic fungal or bacterial middle ear infections spread to inner ear through oval or round window or osteomyelitis of petrous bone. Cytomegalovirus, adenovirus and HSV have been isolated from perilymph with AIDS at death.

**Clinical**
Symptoms vary from dizziness, hearing loss, tinnitus, to ear pain

d. **Chronic infectious / granulomatous**

i. **Syphilis**

Hearing loss (6.5% of unexplained SNHL) occurs in secondary, tertiary and congenital syphilis.

7% patients of Meniere’s disease have syphilis.

Obliterative endarteritis and mononuclear infiltration produce periostitis and gummatous (central necrosis with surrounding lymphocytic infiltration and vascular occlusion) osteitis/periostitis. Narrowing of endolymphatic duct and sac due to atrophy and fibrosis leads to endolymphatic hydrops.

1. **Secondary**
   - Hearing loss (abrupt onset, bilateral, progressive)
   - Minimal vestibular symptoms
   - Patients may have headaches, stiff necks, cranial nerve palsies, optic neuritis, secondary syphilitic rashes, and lymphadenopathy.
   - CSF: Lymphocytic pleocytosis, elevated proteins, and normal glucose
   - Hennebert’s sign (positive fistula test with intact tympanic membrane) may be present.
   - Episodes of acute vertigo are similar to Meniere’s disease.

2. **Tertiary**
   - Otologic features are similar to late congenital syphilis.
   - CSF may have minimal pleocytosis and elevated or normal protein.
   - Electrocochleography: Features similar to Meniere’s disease.

3. **Congenital**

   Early (birth to 3 years)
   - Rapid bilateral profound symmetric SNHL.
   - Relatively few vestibular features, which vary from mild imbalance to protracted vertigo with vegetative features lasting days.
• Meningoneurolabyrinthitis: Systemic features such as meningitis overshadow otologic symptoms.
• Extensive multisystem damage of other organs proves fatal. Late (8 to 20 years)
• Hutchinson's triad, which consists of sensorineural hearing loss (SNHL), interstitial keratitis and notched incisors, is an exclusive feature of congenital syphilis. Hutchinson's teeth (peg-shaped and notched permanent upper central incisors) and mulberry molars (first lower molar grinding surface has many tiny cusps).
• Bilateral, asymmetric, progressive but fluctuating flat SNHL. Speech discrimination scores disproportionately low to pure tone threshold. Recruitment present.
• Tinnitus may appear intermittently.
• Episodes of acute vertigo are similar to Meniere's disease.
• Hennebert's sign (positive fistula test with intact tympanic membrane due to softened gummatous otic capsule) may be present.
• Tullio phenomenon (nystagmus and vertigo caused by loud noise) is often seen.
• Caloric responses are often decreased.
• Interstitial keratitis occurs in 90% cases.

Diagnosis:
• Treponemal tests such as Free Treponemal Antigen Absorption (FTA-abs) and Microhemagglutination Treponema Pallidum (MHATP), which detect organism are more sensitive (95%) than non-treponemal tests screening VDRL and RPR, which detect only 70% patients of otosyphilis.
• Both positive serum rapid plasma reagin (RPR) test and positive fluorescent treponemal antibody-absorption (FTA-ABS) test or positive CSF-Venereal Disease Research Laboratory (CSF-VDRL) test. LP is usually not required as otosyphilis is occasionally associated with neurosyphilis.
• Identification of spirochetes by dark field examination of perilymph obtained by stapes footplate labyrinthotomy.

Prognosis
It is poor in early congenital and better in others. Hearing improvement especially speech discrimination occurs in 35% to 50% of patients.

ii. Lyme disease (Bannwarth's Syndrome)
Multisystem spirochete (Borrelia burgdorferi) disease, which involves skin, nervous system, heart, and joints.
Transmitted by Ixodes ticks from the primary reservoirs (white-footed mice and white-tailed deer)
In USA, initially recognized in 1975 in Lyme, Connecticut. In Europe it is called Bannwarth's Syndrome. There are 3 clinical stages of disease. Affects all ages and both sexes.

Otological features
• Acute Facial nerve palsy - Usually unilateral and recovers completely and spontaneously after weeks to few months.
• There may be preceding H/O ear pain, facial pain or paresthesias.
• Intensely red and violet nodules (lymphocytoma) on the ear lobe.
• Sudden SNHL, positional vertigo and Meniere’s like symptoms have been reported.

**Diagnosis**
- Clinical in endemic regions
- ELISA and Western blotting: Detection of specific antibody to *B. burgdorferi*

**iii. Tuberculosis (TB)**

HIV and emerging resistance to anti-TB drugs have led to rise in incidence of TB. *Mycobacterium tuberculosis* is most common. Atypical mycobacteria include *Mycobacterium avium* and *Mycobacterium fortuitum.* Isolated middle ear and mastoid involvement without pulmonary disease though rare may occur.

Spread: Hematogenous, lymphatic, Eustachian tube, direct through TM perforation.

**Clinical**
- TB OM: Thickening tympanic membrane (TM) and middle ear (ME) mucosa, intractable ME Effusion, Conductive HL, and destruction of ossicles. No pain and tenderness.
- Seropurulent otorrhea
- Multiple perforations, which coalesce and cause loss of total TM; Myringotomy site may enlarge progressively. ME mucosa become hyperemic with polypoid granulations.
- Sequestration of bone, destruction of inner bone, facial nerve, mastoid tip (nontender cold Bezold’s abscess)
- Auditory and vestibular function may be first to manifest and mimic chronic otitis media (COM).
- Nontender upper deep cervical lymphadenopathy.

**Diagnosis**
- Biopsy from ME/Mastoid: Granulomatous process with multinucleated giant cells (Langhans’ cells), AFB TB
- Culture and sensitivity (takes weeks): For accurate identification of mycobacterial species and drug-resistant isolates.

**DD**
- Wegner’s Granulomatosis: Skin tests for TB and demonstration of AFB absent. Presence of antineutrophil cytoplasmic antibodies (ANCAs)
- Langerhans Cell Histiocytosis (LCH)
The disease, which exact etiology and pathogenesis are not known, is characterized by proliferation of cytologically benign histiocytes.

**Otological Features:**

- LCH often mimics chronic suppurative otitis media (CSOM), which does not respond to routine antibiotic treatment and mastoid surgery.
- LCH must be suspected in cases of bilateral destructive ear disease, an elevated ESR, exuberant granulation tissue after mastoid surgery, persistent discharge and associated skin and systemic lesions.
- Mastoid is a common site and may be the initial presentation of LCH.
- Erosion / secondary infection of posterior bony EAC wall, mastoid, zygomatic, or squamosal portions.
- SNHL, vertigo and VII CN palsy occur occasionally.
- The most common symptom is otorrhea, which may be followed by nontender postauricular swelling, hearing loss, and vertigo.
- Most common sign: Granulations and polyp in EAC.
- Tympanic membrane perforation, otitis media and externa, fistula between mastoid and EAC.
- Positive fistula test with intact TM occasionally.
- Diagnosis: Radiographs and biopsy. Positive immunostaining for S100 protein and electron microscopy (Birbeck granules, expression of CD1 on cell surface) will confirm the diagnosis.

**Types:**

- **Unifocal Eosinophilic Granuloma**
  A solitary osteolytic lesion in femora, pelvis, scapulae, vertebrae, ribs, mandible, maxilla, temporal bone or any other skull bone. May cause local pain or pathological # in children and young adults.
- **Hand-Schuller-Christian disease**
  Multifocal osteolytic lesions occur in children of less than 5 years old.
  The disorder also involves skin, viscera and lymph nodes.
  Multiple lesions develop within 6 months.
  **Clinical**
  - Fever, anorexia, recurrent upper respiratory tract infections (URI), anterior cervical lymphadenopathy, otitis media and hepatosplenomegaly.
  - Triad of osteolytic skull lesions, exophthalmos and diabetes insipidus is present in 25% of cases.
  - X-ray chest may show diffuse pulmonary infiltration.
  - Diagnosis by biopsy
- **Letterer-Siwe disease**
  This disseminated disorder occurs in children of less than 3 years of age. This virulent disease has high mortality rate.
  **Clinical**
Fever, seborrheic or eczema like rash, oral lesions, lymphadenopathy, hepatosplenomegaly, multiple bony lesions, diffuse replacement of marrow with resulting blood dyscrasias, and pulmonary infiltration with respiratory failure.

**Diagnosis**
1. Radiographs: Osteolytic lesions.
2. Biopsy: Langerhans cells (nuclei deeply indented and elongated; cytoplasm pale and in abundance) with variable number of eosinophils, macrophages, and lymphocytes.

**v. Wegner's Granulomatosis (WG)**

Though considered autoimmune the exact etiology and pathogenesis are unknown.

This granulomatous inflammatory lesion with necrotizing vasculitis primarily affects respiratory tracts and kidneys.

Usually affects 3rd and 4th decades of life in either sex

**Clinical**
- Common presenting features (in 75% to 90% of cases): headache, sinusitis, rhinorrhea, otitis media, fever and arthralgia.
- Pulmonary features (in 65-75% of cases): Cough, pleuritic chest pain, and hemoptysis.
- Kidney (in 60 to 75% cases): Glomerulonephritis
- Eye (15-50%): Conjunctivitis, iritis, scleritis, proptosis
- Dermatologic: Necrotic ulcers, vesicles, or petechiae.
- Otologic: May be the only and initial presenting features. Serous otitis media and conductive deafness are more common than SNHL. Other features include CSOM, granulomatous lesions in middle ear and mastoid may involve facial nerve and inner ear and manifest with rapidly progressive SNHL and loss of vestibular functions.

**Diagnosis**
- X-Ray Chest: Nodular or cavitary infiltrates
- Normochromic and normocytic anemia, thrombocytosis, elevated ESR
- Positive rheumatoid factor, hyperglobulinemia (especially IgA)
- Positive ANCA test. The specificity of positive proteinase 3-specific cytoplasmic ANCA (c-ANCA) in WG is 95%.
- Biopsy: Granulomatous inflammatory lesion with necrotizing vasculitis, multinucleated giant cells and microabscesses.

**vi. Sarcoidosis**

Chronic multisystem disease of unknown etiology.

It most frequently affects lungs and occurs in 3rd to 4th decade of life.

Blacks are 10 times more affected than whites.

**Clinical**
- The most common features include cough, granulomatous skin rashes and bilateral hilar adenopathy.
• The triad of uveoparotid fever (Heerfordt’s syndrome) consists of parotitis, uveitis, facial nerve palsy and mild fever.
• Eye: Iridocyclitis, keratoconjunctivitis
• Lymphadenopathy and hepatosplenomegaly
• Myalgia and arthralgia
• Cardiac failure
• Nervous system: Peripheral mononeuritis or polyneuritis,
• Otologic: SNHL, VII CN palsy (often bilateral, sudden and resolve spontaneously), vestibular dysfunction and occasionally granulomatous lesion EAC, middle ear and mastoid.

Investigations
• X ray chest: Hilar adenopathy
• Hypercalcaemia, elevated serum angiotensin converting enzyme.
• Biopsy: Noncaseating epithelioid granulomas

2. TRAUMATIC
Vertigo and ataxia are common from injuries of head, neck, vestibule and CNS. Vestibular injury can be caused from blunt concussive trauma, penetrating trauma, explosive blast, and barotraumas.

a. Nonpenetrating Trauma
i. Labyrinthine Concussion\textsuperscript{209, 298}

No injury to otic capsule or intralabyrinthine limiting membranes.

Mode of trauma
1. Blunt Head trauma
2. Inner ear barotraumas

Pathology
• Intralabyrinthine hemorrhage and exudation of fluid into endolymph and perilymph (serous labyrinthitis) produce the symptoms.
• Resolution of serous labyrinthitis leads to resolution of symptoms.
• In some cases fibrosis and ossification gradually destroys membranous labyrinth and lead to progressive hearing loss and persistent dizziness.
• Disassociated rotation of brain about the axis of brainstem within the skull in cases of blunt head trauma with a bat can cause hemorrhage, contusion, partial laceration and total resection of VIII CN, which can result in transient or permanent loss.

Clinical
• Both auditory and vestibular complaints (Dizziness, deafness and tinnitus) are short lived and gradually subside over a period of days to weeks. In some cases symptoms worsen and persists over years.
• Auditory symptoms (hearing loss and tinnitus) occur often.
• Dizziness variable and includes mild vertigo, imbalance, visual confusion, and vegetative symptoms (nausea and vomiting). Dizziness, which usually subsides gradually over days to weeks, may persist for years and mimic BPPV or bouts of movement-associated imbalance and Meniere’s disease or episodes of severe vertigo with nausea and vomiting (similar to delayed endolymphatic hydrops).
Investigations

- Audiogram: Loss most apparent at 4 kHz similar to noise-induced HL. HL can be profound to total loss.
- Nystagmus: Acute nystagmus is toward the side of lesion and after hours, changes to contralateral direction.
- Past pointing and falling towards the slow phase of nystagmus.
- Caloric response reduced
- Vestibulo-ocular reflex: Abnormal

ii. Blast Trauma

Explosive blasts can produce >200 dB sound pressure level (SPL).

Mode of injury

- An open handed slap to the ear
- Actual Explosions

Clinical

- Perforation of Tympanic Membrane,
- Ossicular Disruption,
- Inner Ear Damage (more severe when conductive system is not disrupted).
- Hearing loss can be immediate, permanent and profound. High frequency HL recovers spontaneously.
- Vestibular damage is infrequent.

b. Penetrating Trauma (Violation of otic capsule)

Vertigo with vegetative symptoms and SNHL (vary from mild and transient to profound and permanent) heralds labyrinthine damage. Vertigo usually subsides gradually over days to weeks. Nystagmus beats toward normal side. Fall and past pointing are in the direction of the affected ear (toward the slow phase of nystagmus)

i. Temporal bone fracture

(Both otic capsule sparing and disrupting)

1. Transverse (Usually Otic capsule disrupting) – Blows to occipital region. Involves foramen magnum, petrous pyramid and otic capsule, jugular foramen, internal auditory canal, foramen lacerum.
   a. Rupture of membranous labyrinth – Acute vertigo, SNHL, nystagmus beats towards healthy ear. Fall and past pointing towards affected ear. Vertigo gradually subsides over days to weeks.
   b. Laceration of VIII cranial nerve
   c. Facial Nerve paralysis
   d. CSF fistula
   e. Greater risk of intracranial injuries
   f. Higher risk of delayed meningitis because otic capsule enchondral bone does not remodel and heal.

2. Longitudinal (Usually otic capsule sparing) - Blow to the temporoparietal region. Involves squamosal portion, posterosuperior wall of EAC, mastoid air cells, middle ear
and tegmen tympani and mastoideum and tegmen of facial hiatus.

a. Conductive or mixed HL
b. EAC: Note # along the roof, CSF otorrhea, degree of hemorrhage, presence of brain herniation
c. Labyrinthine concussion
d. BPPV

3. Oblique / Mixed

Evaluation

- Secure airway, control bleeding, examine neurological status, stabilize and evaluate cervical spine.
- Assess facial nerve function at the earliest.
- Auricle: Lacerations and hematomas (untreated – chondropathy or cauliflower ear)
- EAC: Note # along the scutum and roof, CSF otorrhea, degree of hemorrhage, presence of brain herniation. Blood and cerumen may be aspirated but never irrigated.
- TM perforations, hemotympanum.
- Ear packing is discouraged and done only in cases of significant bleeding. Profuse bleeding not controlled by packing is managed by carotid ligation or angiography for balloon occlusion.
- Early ear canal stenting, which is required in cases of severely traumatized ear canals, is not done acutely.
- Vertigo and nystagmus – peripheral or central. BPPV common. Vertigo usually resolves spontaneously.
- Fistula test is positive but not done in acute setting (for the fear of introduction of air and infection in the ear). In cases of CSF fistula, indicates perilymph fistula that present with vertigo and nystagmus for more than a week and fluctuating or progressive SNHL.
- Audiogram: In cases of CSF fistula and facial palsy.
- CT Head to assess intracranial hemorrhage
- HRCT Temporal bone: In cases of facial palsy, CSF leak, # roof of EAC, suspected vascular injury.

ii. Introduction of foreign bodies

1. Hairpin
2. Bullet
   Injury to tympanic membrane, middle ear structures, inner ear (subluxation of stapes into vestibule).

iii. Ear Surgery – Stapedectomy

c. Barotrauma

During the occupations of divers, submariners, bridge builders, and pilots, visual and proprioceptive cues are less effective. So the acute vestibular dysfunction (inner ear trauma as a consequence of alteration in atmospheric pressure) can be devastating.

i. Alternobaric trauma

Incidence:
26% divers and 10% to 17% pilots experience alternobaric-like vertigo

*Cause:*
Elevated or asymmetric middle ear pressure and decreasing ambient pressure *during the ascent* in divers and pilots and fliers. Vertigo is relieved by equilibration ME/ambient pressure differences and repressurization (descending several meters)

*Pathogenesis*
Not known. Pressure differences between ME and intracranial/intralabyrinthine space is presumed to play a role.

*Predisposing factors:*
Alteration of Eustachian tube patency during URT infection (rhinitis, sinusitis)

*Clinical*
- Alternobaric Vertigo (Trauma): Transient vestibular and/or auditory dysfunction caused by elevated and asymmetric middle ear pressure between ears. In most cases vertigo, hearing loss and tinnitus resolve in 10 to 15 minutes.
- Nystagmus toward the ear with higher middle ear pressure.

*Preventive measures:*
- Frequent equilibration of ME pressure during diving.
- Topical decongestants
- Avoid diving during URI and sinusitis.

---

**ii. Atmospheric Inner Ear Barotrauma (IEBT)**

*Etiology*
Abrupt pressure differential transmitted into labyrinth such as in divers (both Scuba and breath-holding) and forceful sneezing with closed mouth and nose. Injury to inner ear is usually long lasting or permanent.

Implosive theory: Increased ME pressure displaces oval/round window into labyrinth and can rupture into inner ear.

Explosive theory: Increased intracranial/intralabyrinthine pressure results in outward rupture of oval/round window into ME.

*Pathology*
Varying from labyrinthine concussion, intralabyrinthine membrane tears, to damage to receptors. In some cases oval and round window fistulae occur.

*Clinical*
- Dizziness, SNHL (high frequency), and tinnitus usually *during descent* are most common complaints at depths varying from 10 to 30 feet.
- Vertigo is less common and a rare sole complaint.
- Warning features: Unsteadiness, nystagmus and ataxia.
- Findings of ME barotraumas: TM perforation or hemotympanum
- Audiogram: High frequency HL in 4- to 8-kHz. In some cases total loss.

*Prevention*
- Gentle Valsalva maneuvers at frequent interval during descent.
• Stop descending if ME pressure equilibrium is not achieved.
• IEBT patients once fully recovered should not dive for at least 3 months.
• Patients with permanent loss of either auditory or vestibular functions should avoid diving.

iii. Inner Ear Decompression Sickness (IEDS) and Isobaric Gas Counterdiffusion Sickness (IGCS)\textsuperscript{99,100,132,282,310}
Common due to the use of mixed gas oxyhelium (oxygen and helium) when diving to depths greater than 100 m. Rapid reduction of atmospheric pressure while rapid ascent leads to formation of gas bubble in the body. These occluding gas bubbles in labyrinth and blood vessels damage the inner ear, which is followed by intralabyrinthine fibrosis and new bone formation.

Clinical
• Tinnitus, SNHL, and vertigo in any combination.
• Though other systems are also involved, ear symptoms are most common and are often the sole features of decompression sickness. Vertigo is the presenting complaint and often the sole complaint.
• Symptoms appear during the rapid ascent.

Risk factors
• Rapid ascent after deep and prolonged diving.
• Specified decompression time schedule is not followed.

3. METABOLIC\textsuperscript{36}
   a. Hyperviscosity syndromes
      i. Hyperlipidemia
      ii. Polycythemia
      iii. Macroglobulinemia
      iv. Sickle cell anemia
   b. Diabetes mellitus
   c. Hyperlipoproteinemia
   d. Hypothyroidism
   e. Hormonal fluctuations
      i. Premenstrual
      ii. Perimenopausal
      iii. Oral contraceptive
      iv. Estrogen replacement therapy
   f. Otosclerosis

4. COLLAGEN VASCULAR DISORDERS
   a. Autoimmune Inner-Ear Disease (AIED)\textsuperscript{146,279}

Types:
Primary: Pathology restricted to the ear
Secondary: to other systemic autoimmune diseases and vasculitides.
Primary AIED is rare and less common than sudden sensorineural hearing loss.
Inner ear involvement is rare in multisystemic AIED.

Clinical:
• Rapidly progressive (weeks to months) unilateral to bilateral sensorineural hearing loss. Fluctuations in hearing may occur.
• 50% have vestibular dysfunction and 20% have episodes of vertigo similar to Meniere’s disease.

**DD**
- Sudden SNHL: Unilateral SNHL develops within 72 hours.
- Meniere’s disease: During the first month, the two diseases may be difficult to differentiate as both the conditions can present with fluctuations of hearing and respond well to corticosteroids. Later on more aggressive course of AIED will allow for the differentiation.
- Otosyphilis
- Acoustic Neuroma: May present with sudden or progressive unilateral SNHL.
- Meningitis
- Multiple sclerosis
- Malignancy: Metastatic disease, lymphoma

**Diagnosis:**
Following work up is aimed at detecting evidence of systemic diseases
- H/O recurrent or chronic ocular disease, nephritis, arthritis, pneumonitis, sinusitis, and inflammatory bowel disease
- CBC, DC, ESR
- Rheumatoid factor
- Antinuclear antibody test
- Anti-double-stranded DNA antibodies
- C3 and C4 complement levels
- Raji cell assay for circulating immune complexes
- Fluorescent treponemal antibody absorption test or Treponema pallidum hemagglutination test.
- MRI to rule out cerebellopontine (CP) angle space occupying lesion (SOL).

Primary AIED is difficult to diagnose and following tests may be tried
- Western Blot Assay for Anti-HSP-70 Antibodies and an antibody that binds to 68-kD antigen derived from bovine temporal bone extract.

**b. Susac’s syndrome (Retinocochleocerebral Vasculopathy)**
Microangiopathy of VIII CN, cochlea, retina and brain.

**Clinical**
Vision loss, low frequency HL, vertigo, and neurobehavioral changes.

**c. Cogan’s syndrome (Nonsyphilitic Interstitial Keratitis)**
AIED Cogan’s syndrome is defined by the presence of labyrinthine and ocular pathology.

**Clinical**
- The characteristic features include Meniere’s like hearing loss and vestibular symptoms as well as interstitial keratitis with nonreactive tests for syphilis.
- Hearing loss though initially is peak shaped but later on unlike Meniere’s is bilateral, progressive without spontaneous improvement and frequently becoming profound.
- Labyrinthine features (sudden attacks of vertigo with ataxia and vegetative symptoms) may be coincident with ocular manifestations or may occur up to 6 months before or after the onset of ocular manifestations. Progressive
complete loss of vestibular function, manifested by ataxia and oscillopsia is common.

- Onset is sudden but resolution is gradual. Recurrence is known for years.
- Ocular complaints (infiltration of cornea with lymphocytes, plasma cells, and neovascularization) are photophobia, blurred vision, lacrimation, or pain.
- Patients often have URI within 7 to 10 days of ear and eye features.

**Etiology**

Raised IgG and IgM titer for chlamydia sp during acute stage decrease with remission of Cogan’s. Perhaps Chlamydia infection sensitizes immune system to "self".

Features supporting autoimmune etiology
- Lymphocytes and plasma cells in cornea and inner ear.
- Associate incidence of rheumatologic disorders
- Response to steroids

**Types**

Typical form has interstitial keratitis while Atypical form present with uveitis, episcleritis, iritis or conjunctivitis.

Vertigo, bilateral progressive hearing loss and interstitial keratitis / preceding H/O upper respiratory tract infection such as cold and cough.

1. **Cogan’s syndrome - Atypical form (10%)**—ocular findings of scleritis, episcleritis, corneal neovascularization, papilledema, and retinal detachment.

Systemic features arise from systemic vasculitis such as polyarteritis nodosa, arthritis, glomerulonephritis, gastrointestinal problems. Patients usually have ocular complaints such as photophobia, blurred vision, lacrimation, or pain.

2. **Cogan’s syndrome - Typical form (90%)**—Systemic involvement (mainly heart and lungs includes aortitis, aortic insufficiency, pleuritis, pericardial effusion, coronary arteritis, and myocardial infarction. Patients usually present with cough, breathlessness, chest discomfort such as pain or tightness.

**d. Vogt-Koyanagi-Harada Syndrome**

In addition to symptoms of Cogan’s syndrome, patient has depigmentation of the hair and skin around the eyelashes, loss of eyelashes, and onset of aseptic meningitis.

**e. Wegner’s Granulomatosis**

Granuloma formation and vasculitis initially affect upper- and lower-respiratory tracts and later progresses to other systems and renal involvement. Chronic otitis media and conductive deafness are more common than SNHL (10% cases).

5. **ISCHEMIC**

**Labyrinthine Infarction**—Occlusion of labyrinthine artery alone or one of its branches results in sudden profound permanent cochlear and vestibular loss in the absence of any other neurological features. Vertigo and imbalance gradually improve due to central compensation. In presence of vascular risk factors and H/O TIA or strokes, acute isolated vertigo cases should be evaluated for vascular etiology.

**Small labyrinthine vessels**—Isolated infarction of vestibular labyrinth

**DD**
- Idiopathic Sudden Sensorineural Hearing Loss (SNHL)
• Multiple sclerosis
• Lyme Disease
• Acoustic Neuroma: Uncommon presentation

Vascular Compression of CN8 Complex
• Brief (5-10 seconds) periods of vertigo induced by certain head movements

6. DRUG-INDUCED

a. Aminoglycoside Antibiotics

Aminoglycosides (Streptomycin injections, kanamycin, tobramycin, amikacin, netilmicin, and sisomycin, gentamycin injections or ear drops, neomycin ear drops). They can damage kidneys and inner ear. Though some are more toxic to either cochlea or the vestibule, their toxicity is not selective. Ototoxicity usually occurs after days or weeks of exposure. Auditory toxicity occurs in 20%, while vestibular may occur in 15%. Measurement of peak and trough serum levels of aminoglycosides help in monitoring ototoxicity, which may continue even after cessation of therapy. Audiometric monitoring (especially high-frequency audiometry) is done pretreatment, post-treatment and weekly in cases of long treatment. Measurement of peak and trough serum levels of gentamycin help in monitoring nephrotoxicity, however, ototoxicity can develop when all of these are controlled.

Histopathology:
Organ of Corti
• Damage to outer hair cells of basal turn, which spread to apical region.
• Progressive destruction of spiral ganglion cells.
• Thinning of stria vascularis

Vestibular System
• Hair cell damage in apex of cristae and in striola regions of maculi and may extend to type I hair cells in the periphery of vestibular sensory system.

Clinical
• High frequency hearing loss is detectable, which progresses to lower frequencies and speech range.
• Onset of vestibular ototoxicity is unpredictable. Imbalance and ataxia are worsened by motion or ambulation. May lead to oscillopsia and complete inability to walk without assistance. Clinical improvement, which is rarely complete, may begin after 2 months.

Risk Factors
• Bacterimia and fever
• Hepatic and renal dysfunction.
• Combination of other ototoxic drugs – Cisplatin, Ethacrynic acid and Furosemide, Amphotericin-B, and cyclosporin
• Prior exposure to aminoglycoside.
• Genetic predisposition: Mitochondrial RNA mutation may sensitize the auditory system even to single dose of drug.

b. Antineoplastic agents – Cisplatin
• Indications: Carcinomas of ovary, testis, bladder, lung, head and neck.
• Side effects: Nausea, vomiting, neurotoxicity, ototoxicity and nephrotoxicity.
- Hearing loss is related to dose, age, noise exposure, low serum albumin, anemia, other ototoxic drugs and cranial irradiation.

**Clinical**
- Hearing loss (permanent and bilaterally symmetric), ear pain, or tinnitus (transient or permanent). High frequencies are affected first and may extend into middle frequencies range.
- Vertigo and disequilibrium especially in patients with preexisting vestibular problems.
- Damage to vestibular hair cells
- Audiometric monitoring (especially high-frequency audiometry) before beginning of each cycle
c. **Anticoagulants:** Vertigo and hemorrhage into inner ear or brain
d. **Loop diuretics:** Furosemide
SNHL (temporary or permanent) may be accompanied by tinnitus and vertigo.
e. **Quinine toxicity (Cinchonism)**
- Deafness (reversible or permanent), vertigo, tinnitus, headache, visual loss and nausea
- Audiogram: High frequency loss, 4-kHz notch, speech discrimination less than 30%.
f. **Erythromycin**
- Transient blowing tinnitus and loss of hearing (usually flat type of SNHL) and in some cases vertigo. Common in elderly and impaired kidney or liver function and other ototoxic drugs.
- Some patients have confusion, fear, psychiatric disturbances, visual changes, slurred speech, sensation of being drugged, or lack of control.

7. **ENDOLYMPHATIC HYDROPS** — Vertigo Lasting Minutes to Hours
   - Idiopathic (Meniere’s)
   - Secondary
     - Otic syphilis
     - Delayed endolymphatic hydrops
     - Cogan’s disease
     - Recurrent vestibulopathy
   - **Meniere’s Disease (Idiopathic Endolymphatic Hydrops)**
     - Symptom complex consists of spontaneous episodic vertigo, fluctuating SNHL, tinnitus, and often a sensation of fluctuating ear fullness.

**Etiology**
The causative roles of HSV, CMV, or VZV remain uncertain.
Familial occurrence in 10% to 20% cases. Autosomal dominant mode of inheritance.
Autoimmune: Certain genetically acquired major histocompatibility complexes (MHC) specifically human leukocyte antigens (HLA) B8/DR3 and Cw7 have been associated with Meniere’s disease.
Allergic and immunology

**Pathogenesis**
Over accumulation of endolymph at the expense of perilymphatic space results in the distortion of membranous labyrinth (Alterations in the size of endolymphatic duct and sac along with reductions in tubular specializations of
the lining of these structures). Perisaccular ischemia and fibrosis leads to inadequate absorption of endolymph by the endolymphatic sac. Endolymphatic hydrops mainly occurs in pars inferior (cochlea and saccule) and changes in pars superior (utricle and SCC) are usually less obvious. The distended Reissner's membrane can obliterate the scala vestibuli. Saccular distension can distort not only utricle and SCCs but can also come in contact with stapes footplate, which can cause Hennebert's sign. Hair cells and their neurons are usually spared. Membranous rupture, which can occur in any part of the inner ear, allows leakage of potassium-rich endolymph into perilymph. The high concentration of K depolarizes the neurons and inactivates both vestibular and auditory neurons that result in vertigo (paralytic nystagmus) and deafness. Healing of membranes allows restitution of normal chemical and clinical status. Repeated membranous rupture and potassium exposure lead to chronic deterioration in the functions inner ear.

CT scan: Hypoplasia of the endolymphatic sac and duct results in decreased visualization of vestibular aqueduct and reduction in periaqueductal pneumatization. Reduced retrolabyrinthine bone might be predisposing factor to disease.

MRI: Significantly smaller and shorter endolymph drainage system.
Gadolinium-enhanced MRI: Enhancement of endolymphatic sac reflects inflammation of sac.

Incidence
Varies from 7.5 to 157 per 100,000 persons.
Affect primarily Caucasians with a slight female preponderance.
About 10% of dizziness patients have Meniere's disease (MD).
Age of onset: 4 to 90 years but peak incidence are in 40- to 60-year age group.
Bilateral disease develops in 47% of cases followed up for 20 years.

Clinical
- The typical presentation consists of recurring attacks (3-11 per year) of spinning vertigo (96.2%) in horizontal axis (for minutes to hours usually 2 to 3 hours) with tinnitus (91.1%) and ipsilateral hearing loss (87.7%).
- Attack lasting more than a day is inconsistent with the diagnosis.
- Attacks are usually preceded by an aura consisting of fullness in the ear, increasing tinnitus and decrease in hearing. Attacks may be sudden without any warning or may awaken patient from sleep.
- Attacks may cease spontaneously after 2 years (57%), 8.3 years (71%) or may occurs for 20- to 40-years.
- In the early phases of disease patient may have either vestibular (recurrent vestibulopathy) or auditory symptoms. The terms vestibular Meniere or Cochlear Meniere are considered inappropriate by AAO-HNS Committee on Hearing and Equilibrium.
- Cluster of attacks separated by long remissions. Single, sporadic attacks or periods of unrelenting recurring attacks. Patient may be minimally inconvenienced or completely incapacitated. Emotional impact may be equivalent to major medical problem.
- Attacks are often accompanied by nausea, vomiting, diarrhea, or sweating. Vertigo is exacerbated with any head movement.
- Between the attacks patient may be totally symptom free or feel disequilibrium, lightheadedness or tilt.

**Drop attacks (Otolithic crises of Tumarkin)**
- Occasionally sudden unexplained falls without loss of consciousness or associated vertigo. Patient feels pushing or moving during this short-lived spell.
- Acute utriculosaccular dysfunction leads to inappropriate postural adjustment via vestibulospinal pathway.
- Other causes of drop spells: Cardiogenic vertebral basilar insufficiency and migraine.

**Hearing loss**
- Fluctuating and progressive SNHL.
- Diplacusis (a difference in the perception of pitch between the two ears).
- Intolerance to loud sounds (recruitment).

**Tinnitus:**
- Nonpulsatile, whistling or roaring, continuous or intermittent.
- Become louder or changes pitch as the attack approaches and improves after the attack.

**Lemoyez Attacks**
Increased tinnitus and hearing loss precede the vertiginous episode and dramatically resolve with onset of vertigo.

**Nystagmus**
- The direction of horizontal nystagmus, which is the cardinal finding, varies over the course of the attack. So the involved ear cannot be determined just on the basis of direction of nystagmus.
  - Early irritative nystagmus: ipsilateral
  - Later paralytic nystagmus: contralateral
  - Late recovery nystagmus: ipsilateral

**Audiogram**:
- Low frequency fluctuating and progressive sensorineural hearing loss and a coincident nonchanging high frequency loss. Average pure tone loss of 50 dB. Profound deafness is rare.
  - Peaked at 2 kHz (tent-like audiogram).
  - Overtime hearing loss flattens.
  - Recruitment present in 56% cases.
  - A mean speech discrimination score of 53%.

**Investigations**
Electronystagmography: Caloric response
- Significantly reduced in 48% to 73.5% cases.
- Complete loss in 6-11%.

**Dehydrating Agents**
Urea, glycerol, and furosemide produce measurable improvement in audiometric score, reduction in summating potential negativity (electrocochleography) and change in gain of vestibulo-ocular response to rotational stimulation. Sensitivity and specificity vary widely.

**Glycerol Test**
- Glycerin 1.5 g/kg mixed with equal volume of juice followed by serial audiograms over 3 hours.
Positive test: 25 dB shift at 3 consecutive frequencies, or 16% improvement in speech discrimination.

**Electrocochleography**
- Infrequently used.
- Summating potential (SP): Larger and more negative (distension of basilar membrane into scala tympani)
- Action potential (AP) of CN8: Reduction in amplitude
- SP/AP ratio: Increases (Most commonly used value)

**DD**
- Basilar migraine
- Autoimmune disease of inner ear
- Syphilis
- Cardiogenic Vertebral Basilar Insufficiency
- Migraine.

**b. Delayed Endolymphatic Hydrops**
Meniere’s like attack of vertigo in cases of preexisting profound loss of hearing in one or both ears (1-74 years) due to trauma (physical and acoustic), idiopathic sudden SNHL, labyrinthitis (mumps, influenza, mastoiditis, meningitis, diphtheria, measles) and idiopathic childhood deafness.

Lasix test: Dehydrating agents such as furosemide (Lasix) improve the vestibular response of the affected ear.

**c. Recurrent Vestibulopathy**
Relatively common. Formerly called Vestibular Meniere’s.
Cause not known but some postulates viral due to similarities with vestibular neuronitis.

**Clinical**
- Mean age of onset 37 years with equal sex distribution.
- Attacks of vertigo (sudden, lasting minutes to hours and recurring at variable interval) similar to Meniere’s disease without audiologic symptoms.
- 9% patients have additional features of BPPV.
- Diagnosis may change to Meniere’s disease as about 14% patients develop audiologic symptoms.
- No neurological features
- Caloric response is reduced only in 22% (50% in Meniere’s)
- 70% patients recover completely over 9.5 years.

**8. NEOPLASTIC**

**a. Cerebellopontine Angle Tumors**
- Causes disequilibrium or unsteadiness rather than true vertigo because central compensation occurs over time due to slow growing nature of tumor. Disequilibrium and positional vertigo may occur because of cerebellar compression.
- The most common CP angle tumor is Acoustic neuroma and rare tumors include Meningioma, Epidermoid, Nonacoustic neuroma, Paraganglioma, Arachnoid cyst, Hemangioma. Other very uncommon tumors are Metastatic, lipoma, teratoma, chordoma, chondrosarcoma, and giant cell tumor.

**b. Multiple Myeloma**
Malignancy of plasma cells (multiple plasma cell tumors) derived from B lymphocytes. Median age of onset is 60 years.

**Clinical**
- Severe bone pain, pathological fractures, renal and bone marrow failure, hypercalcaemia, recurrent infections.
- Otological symptoms, which may occasionally be presenting features, are usually overshadowed by features of diffuse disease.

**Diagnosis**
- Electrophoresis: M component (Monoclonal protein) in serum and urine
- CBC: Normochromic and normocytic anemia
- Hypocalcaemia,
- Elevate BUN
- X ray skull lateral view/ CT scan: Punched-out osteolytic lesions.
- Bone marrow aspirations: Infiltration by plasma cells.

**Plasmacytoma**
- Only one plasma cell tumor without marrow plasmacytosis can occur in temporal bone (solitary bone plasmacytoma) or soft tissue (extramedullary plasmacytoma).
  - It can affect younger persons and has an indolent course (10 or more years).
  - M component is present in < 30% of cases.
  - Radiographs/CT scan: Rounded lytic lesions

**c. Leukemia**
Submucosal Leukemic infiltration, hemorrhage, and secondary infection occur in TM, ME and mastoid.

**Clinical**
- Acute and chronic SOM, ME effusion, thickening of TM, conductive and SNHL, sudden SNHL, vertigo, facial palsy, and skin lesions in pinna or EAC.

**Granulocytic Sarcoma or Chloroma of Temporal Bone**
Localized extramedullary tumor (immature myeloid cells) related to myelogenous leukemia.
Otologic features can manifest during initial presentation.

**d. Temporal Bone Tumors**

**i. Carcinoma of EAC and Temporal Bone**
EAC: Squamous cell carcinoma, basal cell carcinoma and adenoid cystic carcinoma (in order of decreasing frequency).
- Usually affects 40 to 60 years old persons.
- Patients usually present as cases of CSOM or chronic OE.
- Biopsy should be taken in cases of intractable case of chronic OE.
- Persistent and inordinate pain in EAC should raise the suspicion.
- Meaty or polypoid mass in EAC (Squamous cell carcinoma).
- Small pimple with significant pain (adenoid cystic carcinoma).
- Serpiginous ulceration (basal cell carcinoma).
- Labyrinthine involvement causes SNHL and vertigo.
- Late features: Parotid mass, CN palsy, cervical lymphadenopathy.
- Tumor may originate from nasopharynx.
ii. Metastatic Malignant Tumors\textsuperscript{119}
- Primaries: Breast, lung, prostate, and skin (in order of decreasing frequency)
- Spread: Hematogenous
- Usually osteolytic but some time osteoblastic (prostate or breast) occurs in petrous apex and IAC of temporal bone.
- Otic capsule though relatively resistant to neoplastic invasion can also get involved (SNHL, vertigo, facial palsy).
- Involvement of EAC, ME or E. tube may cause pain and conductive HL.
- Otologic manifestations can occasionally present as first symptoms of malignancy.

**Meningeal Carcinomatosis**
- Unilateral SNHL may present as CP angle tumor.
- Bilateral SNHL may present as immune-mediated inner-ear disease.
- Diagnosis: CSF cytology.

9. MISCELLANEOUS

a. **Bilateral Vestibular Hypofunction**\textsuperscript{57, 102, 172, 234, 286, 327}

*Causes*
- Ototoxic drugs
- Degenerative diseases of cerebellum
- Meningitis
- Systemic autoimmune diseases
- Trauma
- Bilateral Meniere’s disease
- Idiopathic – 20% cases

*Clinical*
- Oscillopsia with head movement
- Varying severity in disturbances of gait
- In cases of Injection streptomycin, oscillopsia and disequilibrium begin abruptly and worsen over 2 to 3 days. Even pulse produces enough head movement to cause oscillopsia and difficulty in reading.
- Gentamycin: 3% patients develop vestibular damage. Gait ataxia most common manifestation. Dynamic visual acuity (reading paper while moving head horizontally at a frequency of about 2 Hz), Romberg, head thrust tests and assessment of gait while making head movement should be done daily.
- Balance improves as patient learns how to use visual and somatosensory information to make up for vestibular loss. Spontaneous recovery occurs in many cases but prolonged disability is not uncommon.

b. **Familial Vestibulopathy**\textsuperscript{19}
- A rare syndrome
- Autosomal dominant inheritance.

*Clinical*
- Sudden attacks of vertigo followed by chronic disequilibrium
- Vertigo lasts for minutes followed by several hours of subsequent
disequilibrium, which may become chronic with associated oscillopsia
due to progressive vestibular insufficiency.
- The spells recur at variable intervals for many years. Stress is the
trigger in some cases.
- Auditory symptoms such as deafness and tinnitus are absent.
- All patients have H/O Migraine headache, which is not associated with
spells of vertigo.

**Investigations**
- Imaging normal.
- Features of damage to bilateral peripheral vestibular organ:
  - Caloric tests: Hypoactive to no response
  - Spells of vertigo successfully terminated with acetazolamide, which is
    presumed to reduce the acidity of brain.

**Other variants**

**Vertigo, Migraine Headache and Essential Tremors Syndrome**
- Vertigo lasts minutes to hours
- All patients have H/O Migraine headache, which is not associated with
  spells of vertigo.
- Spells of vertigo triggered by stress, exercise, and lack of sleep.
- Long term progressive vestibular deficit absent (But present in Familial
  Vestibulopathy).
- Acetazolamide curtails spells of vertigo as well as migraine.

**Hemiplegic Migraine**
- Spells of Hemiplegia, Migraine headache and Vertigo
- Localized to chromosome 19p. Presumably dysfunction of an ion
  channel.
- Acetazolamide effective.

**c. Sudden Sensorineural Hearing Loss (SNHL)**
- Usually patient either awakens in the morning with hearing loss or
  hearing loss develops within minutes to several hours (<3 days).
- Unilateral in 90% cases
- 50% patients have accompanying vertigo or imbalance.
- Recovery, which is usually 50% and related to age (better chances in
  <40 years) and severity (less chances in profound HL), occurs in days to
  weeks. After 1 month little chance of recovery.
- Causes are many. Three hypotheses are: Vascular, Traumatic and Viral
  (mumps and measles).

**DDs:** Otosyphilis, Lyme disease, sarcoidosis, acoustic neuroma,
thromboembolic phenomenon of labyrinthe vessels.

**d. Perilymphatic fistula (Inner ear fistula): Round or oval window
fistula**
Inner ear fistulae provide communication between the perilymphatic space and the middle ear or intramembranous communication between endolymphatic and perilymphatic spaces. Common sites: Oval and round windows

**Etiology**

i. Barotrauma  
ii. Penetrating trauma  
iii. Surgery -- Stapedectomy  
iv. Head trauma  
v. Explosive blast  
vi. Physical exertion (Heavy lifting or straining)  
vii. Congenital  
viii. Spontaneous in cases of congenital hearing loss (Congenital malformations such as Mondini's deformity). Fistula should be ruled out in cases of congenital hearing loss.290

**Clinical**

- Variable auditory and/or vestibular symptoms.  
- Dizziness variable and include episodic incapacitating vertigo like Meniere's, positional vertigo, motion intolerance, to occasional disequilibrium.  
- Hennebert's phenomenon: Disequilibrium after nose blowing and weight lifting (due to increase in CSF pressure)  
- Tullio's phenomenon: Vertigo after exposure to loud noises.  
- Hearing loss is usually present but not must.  
- Audiogram: variable SNHL; (high frequency) HF loss to a (low frequency) LF loss or flat one. Fluctuating pure tone thresholds and speech discrimination scores. Isolated mild conductive HL.  
- Electrocochleography: Larger summating potential (nonspecific)  
- Caloric response: Reduced (nonspecific)  
- Spontaneous inner ear fistula in cases of congenital hearing loss (congenital malformations such as Mondini's deformity).

**Diagnosis**

**Confirm by following tests**

1. Fraser test: Improvement in pure-tone threshold or speech discrimination after the patient has been in Trendelenburg position for 30 minutes.  
2. Fistula Test: conjugate contralateral slow deviation of the eyes followed by 3-4 ipsilaterally directed beats of nystagmus after introducing positive pressure in the ear (by pressing tragus or using pneumatic speculum).  
3. Surgical exploration in cases of positive fistula test.  
4. Fiberoptic exploration with rigid scope through myringotomy or flexible endoscope through Eustachian tube.  
5. Free amino acid content of fluid and CSF/perilymph specific protein tau Transferrin differentiate between perilymph and mucosal secretions.

**DDs**

- Inner ear fistulae mimic
- Meniere’s disease
- Infection Labyrinthitis
- Acoustic Neuroma
- Superior semicircular canal dehiscence syndrome
- CNS lesions such as Chiari malformations (cerebellar tonsils displaced caudally through foramen magnum): Confirm by positional downbeat nystagmus and ocular motor findings localizing to vestibulocerebellum

e. **Benign Paroxysmal Positional Vertigo (BPPV)**

Vertigo Lasting Seconds

**Etiological factors**
- None in 48% cases
- Most common are closed head injury and vestibular neuronitis (vertigo lasting days).
- Advanced age
- Infections
- Surgery (stapedectomy or nonotologic) and
- Prolonged bed rest
- BPPV can develop in cases of Meniere’s and recurrent vestibulopathy.

**Pathogenesis**
- Cupulolithiasis: Deposition of otoconia on the cupula of posterior SCC.
- Canalithiasis: Free floating material (debris) within the lumen of posterior SCC.

**Types**
- Posterior SCC BPPV
- Lateral SCC BPPV
- Superior SCC BPPV

**Incidence**
- Most common cause (20% to 40%) of peripheral vertigo
- Age – 11 to 84 years; mean age of onset 4th to 5th decades. Incidence increases with age.
- Pediatric BPPV patients have association with migraine.
- Slightly increased incidence in females.

**Clinical**
- Sudden brief (seconds) spells of severe vertigo associated with change in head position such as
  - Rolling over in bed
  - Getting into bed and assuming a supine position or
  - Arising from a bending position or
  - Extending the neck or
  - Turning rapidly.
- Vertigo spell lasts for seconds and never more than a minute, though patients usually complains of longer subjective feeling of dizziness.
- Bouts of vertigo are clustered in time. Remissions may last for months or more.
- The active spells may be associated with the feeling of lightheadedness or mild imbalance, which is worsened by head movement.
- Some patients have chronic balance problem, which may be worse at the time of awakening from the sleep.
Diagnosis
Confirm the diagnosis through the observation of classic eye movements (enhanced by the use of Frenzel Lenses) in association with Dix-Hallpike maneuver. The condition is classified active if rotatory nystagmus is elicited by Dix-Hallpike test. Patients with a typical history of BPPV, but a negative Dix-Hallpike test and whose symptoms had settled are diagnosed resolved BPPV.

Dix-Hallpike Maneuver for Posterior Canal BPPV
1. Presence of Latency of onset of nystagmus. Duration <1 minute. Associated with vertigo. Nystagmus disappears with repeated testing (fatigable). Vertigo may recur with nystagmus in the opposite direction on return of head to upright position.
   a. Combined vertical upbeating and rotary (torsional) component beating toward downward eye (superior poles of eyes beat toward the downward ear) – most common findings.
      i. BPPV – Canalithiasis of Posterior semicircular canal –
         Most common
   b. Combined vertical downbeating and rotary (torsional) component beating toward upward eye (superior poles of eyes beat toward the upward ear) – very rare
      i. BPPV – Canalithiasis of Superior semicircular canal –
         Very rare
   c. Pure vertical nystagmus
      i. Not BPPV
      ii. Central vestibular disorder
   d. Nystagmus present with patients head turned to both right and left
      i. Bilateral BPPV
      ii. Head is not positioned correctly in the plane of posterior canal during testing of the normal side.
      iii. Head injury or Brainstem ischemia

2. In head hanging position nystagmus begins without a latent period or persists with a constant slow-phase velocity for as long as the head position is maintained. Subjective vertigo is much less than expected from the intensity of nystagmus - Central Vestibular disorder.
   a. Pure vertical nystagmus usually down beating with respect to the head – most common central type – Other following ocular motor abnormalities are present gaze-evoked nystagmus, impaired smooth pursuit, impaired vestibulo-ocular reflex (VOR) suppression, rebound nystagmus
      i. Multiple sclerosis (MS)
      ii. Cerebellar lesion - Chiari malformations (cerebellar tonsils displaced caudally through foramen magnum)
      iii. Lesion at craniocervical junction

Modified Dix-Hallpike maneuver (Horizontal Positional Nystagmus)
1. Horizontal nystagmus may beat toward (geotropic) or away from (ageotropic) the downward ear. Shorter latency and increases in magnitude while maintaining the test position. Less susceptible to fatigue with repetitive position.
   - Lateral Canal BPPV (17% of cases). The pathogenesis is usually Cupulolithiasis with or without canalolithiasis. Particles are usually in the long arm of the lateral canal (canalolithiasis) either far from ampulla (geotropic nystagmus) or near the ampulla / on the opposite side of cupula (ageotropic nystagmus), floating within the endolymph or embedded in the cupula (cupulolithiasis).
2. Sustained, large amplitude nystagmus, which is present during visual fixation. Nystagmus occurs in more than one head position. Nystagmus has an associated vertical (especially down beating).
   - Central vestibular disorder
   - Alcohol intoxication

**DD**
- Vascular Compression of CN8 complex
- Multiple Sclerosis
- Acoustic Neuroma
- Lesion at cranio-cervical junction - Chiari malformations

**f. Superior Semicircular Canal Dehiscence Syndrome**

Dehiscence (developmental or congenital abnormality) of the bone overlying the Superior Semicircular Canal responds to sound and pressure stimuli. This third mobile window into the inner ear probably causes dissipation of acoustic energy transmitted through air conduction and manifest as apparent conductive hearing loss.

**Clinical**
- Clinical manifestations may be only vestibular or only auditory or both vestibular and auditory.
- Dizziness appear after nose blowing / weight lifting (Hennebert's phenomenon), which increases CSF pressure and/or after exposure to loud noises (Tullio's phenomenon) and/or after straining, increased pressure in ear.
- Vertigo and oscillopsia induced by loud noises or change in intracranial / middle ear pressure. Vertical oscillopsia in bilateral cases.
- Patients may complain of hearing the movement of their eyeball and pulse.
- Nystagmus is vertical and torsional. Positive pressure (loud sounds, increased pressure EAC, Valsalva maneuver against pinched nose) causes ampullofugal deflection that manifest nystagmus directing downward and torsional toward the affected ear. Negative pressure (decreased pressure in EAC, Valsalva against closed glottis and jugular venous compression) causes nystagmus conversely that is fast phase upward torsional fast component toward normal ear.
- Chronic disequilibrium.
- Weber lateralizes to affected ear. Tuning fork may be heard when placed on lateral malleolus of the foot.
- Audiogram: Bone conduction <0 dB. Air-bone gap (greatest at lower frequencies), which exist with normal air conduction threshold, is in the range of 24+-7 dB from 250-4000 Hz.

**Diagnosis**
- HRCT Temporal Bone: 0.5 mm-collimated helical CT scans with reformation of images in the plane of superior canal. The bone overlying the intact superior canal becomes significantly thinner.
- Electrooculography: Loud clicks (110 dB, 100 microsecond duration) evoked vestibulo-ocular reflex. Short latency vertical eye movement in affected ear.

**g. Paget's Disease (Osteitis Deformans)**

**Etiopathology**
- Unknown etiology and postulations include endocrine, metabolic, vascular, autoimmune, neoplastic, viral infection.
- Genetic factor (locus on 18q) inherited as an autosomal-dominant trait with high penetrance.
- Osteolytic and osteoblastic (burnt-out phage) changes occur in axial skeleton. Osteoclastic resorption of marrow containing bone with increased vascularity and fibrous tissue occur.
- Irregular new bone formation and curved cement lines produce typical mosaic appearance.
- In temporal bone remodeling of inactive Pagetic bone into normal-appearing lamellar bone.
- 3% population above 39 years and 11% above 79 years.

**Clinical**
- Men are affected more commonly than women.
- Onset usually occurs in 6th decade and present with enlarging skull, progressive kyphosis, deformities of pelvis, femur and tibia.
- Otological features: Progressive hearing loss (Mixed, SN or rarely conductive) tinnitus and mild vestibular dysfunction.
- Audiogram: Usually mixed hearing loss with descending bone conduction relatively flat air-conduction. Severity more than those of age-matched healthy subjects.

**Diagnosis**
- X ray skull lateral view and CT scan: Thickened skull table, patchy and ill-defined densities, ill-defined cortical margins of inner ear and IAC.
- Differentiating features from Otosclerosis: Later age of onset, mixed and more SNHL, enlarged calvaria, and enlarged and tortuous superficial temporal artery and its anterior branches, elevated serum alkaline phosphatase.

**h. Congenital Cholesteatoma (CC) of Temporal Bone**

Four anatomic groups: ME, perigeniculate area, petrous apex and CP angle.

**Clinical**
- ME: Bulging white mass behind TM, conductive HL. Recurrent OM and fistulization through TM.
- Perigeniculate and petrous: Facial twitching or insidious or rapidly progressive facial nerve palsy.
- SNHL and vestibular dysfunction may occur due to labyrinthine or IAC erosion.

**Diagnosis**

- HR focused CT of TB: Alteration in normal bony structure.
- MRI
  - T1W: CC have slightly higher signal than CSF. High signal of bone marrow fat. ME effusion has low signal.
  - T2W: Moderately high signal of CC. Bone marrow fat signal fades. ME effusion has high signal.

**CENTRAL VESTIBULAR DISORDERS**

1. Vascular
   a. Ischemic Strokes
      Vertigo occurs commonly and may be the only symptom. Signs and symptoms of brainstem dysfunction usually occur depending on the vascular supply to specific region.
      **Causes:**
      - Common causes include atherosclerosis, embolism, and dissection. Atherosclerosis of subclavian, vertebral and basilar arteries causes thrombosis and infarction in the brainstem. Cerebellum is usually spared due to anastomoses. An embolus from heart or aortic arch can get lodged in distal branch and cause isolated cerebellar infarction.
      - Vascular risk factors: Hypertension, diabetes, hyperlipidemia, smoking, prior TIA
      - Uncommon causes: Arteritis and other inflammatory conditions, hypercoagulation disorders, hyperviscosity syndromes
      **Management**
      - Cerebellar infarction: First few days monitoring for swelling, brainstem compression and hydrocephalus as they may require neurosurgical intervention.
      - Emergency evaluation for thrombolysis therapy by physicians or neurologists.

   i. Dissection of Vertebral Artery
      Young patients without any vascular risk factor of ischemia. H/O minor neck trauma, neck manipulation, neck pain, head pain Dissection and lumen stenosis or occlusion causing vertebrobasilar ischemia
      MRI and MRA confirm the diagnosis.

   ii. Subclavian Steal Syndrome
- Stenosis of proximal part of subclavian artery prior to the origin of vertebral artery: Siphoning of the blood into the arm due to retrograde blood flow down the vertebral artery.
- Vertigo exacerbated by exercising the arm
- Signs and symptoms of brain stem ischemia;
- Bruit in axilla or supraclavicular region;
- Difference in pulse and blood pressure between two arms (20mm Hg)

iii. **Vertebral Artery Kinking**

Though rare, neck movements can lead to kinking of vertebral artery and acute vertigo.

iv. **Vertebrobasilar Transient Ischemic Attacks (TIA)**

- Common.
  - TIA symptoms usually last for minutes and completely resolve within 24 hours.
  - Common in elderly.
  - Risk factors such as diabetes, hypertension, smoking, hyperlipidemia.
  - Vertigo lasts more than a minute but less than several hours.
  - Abrupt isolated vertigo is a common presentation (TIAs of labyrinth or brain). Isolated spells of vertigo continuing for more than 3 months usually rule out TIA.
  - Other common accompanied features are visual (diplopia, visual field defects, blindness, and visual illusions and hallucinations) followed by drop-attacks, unsteadiness-incoordination, and weakness in limbs. Confusion, headache, hearing-loss, loss of consciousness, numbness in limbs, dysarthria, tinnitus, perioral numbness are rarely accompanied with episodes of vertigo.
  - Episodes of hemiparesis, quadriplegia, bilateral blindness, or altered consciousness need urgent evaluation for impending basilar artery thrombosis.
  - Vertebrobasilar angiography: The most common site of abnormality is near the anterior inferior cerebellar artery (AICA) takeoff, which affects the blood supply to labyrinth and flocculus.\textsuperscript{101, 124}
  - Isolated episodes of vertigo in elderly patients, who have vascular risk factors, should be evaluated without delay as an OPD patient. But the patients of vertigo episodes accompanied with weakness of limbs, blindness and altered consciousness should be urgently evaluated for impending basilar artery thrombosis.

v. **Lateral Medullary Infarction – Wallenberg’s Syndrome**

*Causes*

1. Occlusion of vertebral artery or posterior inferior cerebellar artery (PICA): Occlusion of vertebral artery rather than PICA itself
   a. Dissection of the vertebral artery in young persons
   b. Hypertension and atherosclerosis in elderly
2. Demyelinating disease - rare
Though the cerebellum is usually spared, damage to spinocerebellar tract in inferior cerebellar peduncle results in ataxia.

Clinical
- Partial syndrome: Only disequilibrium and a tendency to fall to one side.
- Complete syndrome includes vertigo, nausea, vomiting, diplopia, severe gait and ipsilateral limb ataxia; Ipsilateral Horner’s syndrome and facial anesthesia, and contralateral hemianesthesia;
- Bizarre sensations of body and environment.
- Localizing features: Dysphagia, hoarseness and dysphonia; decreased gag, and ipsilateral vocal cord weakness.

Characteristic ocular motor abnormalities
- Lateropulsion: Overshooting (deviation) of eyes to the side of lesion with a blink or closing lids
- Torsi pulsion: Inappropriate torsion of eye with horizontal saccades
- Spontaneous nystagmus: Mixed horizontal and torsional, usually directed toward normal side.
- Smooth pursuit (patient follows slowly moving target, no faster than 20 degrees/sec) abnormalities: Impaired when target moves away from the side of lesion.
- Saccades (Keeping the head still, patient alternately fixate physician’s nose and then finger, which are held at different positions approximately 15 degrees away from primary position): Vertical saccades have inappropriate ipsilateral horizontal component and appear oblique.
- Skew Deviation and Ocular tilt reaction (OTR): Skew deviation with ipsilateral hypotropia, ipsilateral head tilt, cyclodeviation (rolling of superior pole of cornea), and subjective visual vertical deviation.
  - Vertical diplopia: One image above the other when one eye is covered with red glass (Maddox rod)
  - Torsional diplopia: One image is tilted with respect to other.
  - Alternate cover test: Look for vertical corrective movement.

vi. Lateral Pontomedullary Infarction
Occlusion of AICA results in infarction of lateral pontomedullary region, middle cerebellar peduncle, and anterior inferior cerebellum. Occlusion of labyrinthine artery, which usually arises from AICA, results in infarction of cochlea and labyrinth or VIII CN.

Clinical
- Sudden vertigo, nausea, and vomiting and severe unilateral hearing loss are presenting symptoms.
• Ipsilateral facial palsy is common.
• Ataxia, ipsilateral facial anesthesia and Horner’s syndrome, contralateral body anesthesia may occur.
• Dysphagia and dysphonia are absent.
  o Labyrinthine Infarction – Labyrinthine Artery Occlusion - (peripheral vestibular disorder)
  o Vestibular-Masseter Syndrome: Acute vestibular imbalance and unilateral paresis of muscles of mastication caused due to occlusion of small branch of AICA in rostral medulla and caudal pons.

vii. Superior Lateral Pontine Infarction
Due to the involvement of medial lemniscus there is impaired vibration and position sense of contralateral side

viii. Cerebellar Infarction
- Management may need prompt neurosurgical intervention.
  Surgical decompression and ventriculostomy to relieve hydrocephalus may be life saving. Large infarct produces progressive swelling, which compresses brainstem.
- Swelling and herniation of cerebellar tonsils can lead to quadriplegia, coma and death.

Clinical
- Presentation can mimic acute peripheral vestibular disorder.
- Vertigo and vomiting, ipsilateral limb ataxia, Horner’s syndrome and facial hemianesthesia and contralateral body anesthesia.
- Brainstem features help in localizing feeding vessels PICA, AICA, or SCA.
  o SCA: Due to the involvement of medial lemniscus there is impaired vibration and position sense of contralateral side
  o AICA: Unilateral paresis of muscles of mastication
  o PICA: Dysphagia, dysphonia
- Ocular Motor Abnormalities: Saccadic dysmetria is opposite of Wallenberg’s syndrome.
  o Contrapulsion: Contralateral overshooting of saccade and ipsilateral undershooting of saccade.
  o Vertical saccade: Inappropriate horizontal component directed toward healthy side.

Isolated Cerebellar Infarction
Clinical
Vertigo, vomiting and severe gait ataxia (profound gait imbalance) with relatively little limb ataxia.
- Direction-changing or spontaneous downbeat nystagmus, which is not suppressed by visual fixation.
- 25% elderly people suffering from acute isolated vertigo have a cerebellar infarction.
DD

- Acute peripheral vestibulopathy: Unilateral head thrust sign.

b. Cerebellum and Brainstem Hemorrhage

Headache or stiffness of the neck suggests hemorrhage. Sudden vertigo with multiple neurological findings (localizing signs and symptoms) often rapidly progress to coma and death. Pontine (5%) and cerebellar (10%) hemorrhage constitute 15% of all intracerebral hemorrhages.

Causes

Hypertension (most common), vascular malformations, intracranial tumors, anticoagulation, bleeding diathesis, amphetamine or cocaine use, trauma, and hemorrhagic infarction.

Diagnosis

1. Noncontrast CT or MRI: Shows acute intraparenchymal hemorrhage. MRI reveals acute infarction not shown on CT.
2. Coagulation studies: Immediate reversal of anticoagulation if patient is taking any anticoagulant to prevent expansion of hematoma.
3. Control raised intracranial pressure
   - **Pontine hemorrhage**
     - Large hemorrhage presents with Quadriplegia, decerebrate posturing, disturbances of horizontal eye movements, ocular bobbing, and pinpoint pupil and coma.
     - Smaller hemorrhages have less extensive symptoms and patients often recover.
   - **Cerebellar hemorrhage**
     - Often begins with vertigo, headache, vomiting, and an inability to stand or walk.
     - Severe ataxia or dysmetria, gaze-evoked nystagmus, and stiff neck
     - Abducens and facial palsies can occur from hematoma compression.
     - Compression of brainstem due to expanding hematoma can result in rapid coma and death either immediately or after some period of stability.
     - Absence of head thrust sign will differentiate it from acute peripheral vestibular disease.
   - **Medullary hemorrhage**
     - Less common.
     - Most often presents with sudden vertigo, headache, nausea, vomiting, dysphagia, dysarthria, and other medullary signs.

i. Hemorrhage Parietoinsular Cortex or Superior or Medial Temporal Gyri (Vestibular Cortex)

Acute vertigo in absence of nystagmus or other brainstem or cerebellar abnormalities.

c. Migraine

- Most common cause of dizziness in many neurotologic clinics.
- In USA: 18% to 29% of females, 6% to 20% of males and 4% of children.
- Usually begins in first 3 decades and prevalence peaks in 5th decades.
- Family history is often present.

**Pathophysiology**

In this neurovascular disease there occurs neural induced vasodilatation, which manifests as pain and leads to further nerve activation. The depressed neuronal function from accumulation of extracellular potassium occurs due to the dysfunction of an ion channel in the aminergic nuclei (such as serotonin-producing dorsal raphe nucleus or norepinephrin-producing locus ceruleus) within the brainstem or diencephalons that modulates sensory input and exerts neural effects on cranial vessels\(^\text{120}\). The cause of headache is not well understood. Large extracranial vessels, proximal intracranial vessels, or dura mater can produce headache. Abnormal stimulation within the peripheral or central vestibular structures (brainstem, thalamus and parietoinsular vestibular cortex) may result in vertigo. Central ocular motor signs in association with vertigo arise from brainstem dysfunction involving vestibular nuclei\(^\text{87}\).

**Classification International Headache Society (IHS)**\(^\text{258, 277}\)

- Migraine without aura – Most common type (also called sick headache)
- Migraine with aura
  - Typical aura with migraine headache
  - Typical aura with non-migraine headache
  - Typical aura without headache
  - Familial hemiplegic migraine
  - Sporadic hemiplegic migraine
  - Basilar-type migraine

- Childhood periodic syndromes (Common precursors of migraine)
  - Cyclical vomiting
  - Abdominal migraine
  - Benign paroxysmal vertigo of childhood

- Retinal migraine

**Complications**

- Chronic migraine
- Status migrainous
- Persistent aura without infarction
- Migrainous infarction
- Migraine-triggered seizure

**Clinical**

**Headache Features**

- Repeated attacks of headache lasting 4 to 72 hours
- Headaches have any two of the following characteristics
  - Unilateral
  - Pulsating (throbbing)
  - Moderate to severe
  - Aggravated by physical activity and motion
- Headaches have any one of the following associated feature
  - Nausea and/ or vomiting
  - Intolerance to light (photophobia) and noise (phonophobia)
- Usually history, physical and neurological examinations do not suggest organic or systemic metabolic disease but if they suggest such disorders, they must be ruled out with appropriate investigations.
- Migraine without aura
  - Vague premonitory psychological, constitutional, neurologic and autonomic symptoms may precede attack by hours or days.
- Migraine with aura
  - Any one of the following fully reversible aura symptoms (without any motor weakness) that usually develop gradually over 5-20 minutes and last for less than 60 minutes. Headache usually begins as the aura is decreasing.
    - Visual aura (most common):
      - Flicker lights or simple flashes, specks, lines, or geometric forms, shimmering (shine with faint quivering (shake, tremble, vibration) light) or undulations
      - Blind spots (scotomata), or loss of vision (hemianopia);
      - Visual hallucinations:
        - Fortification spectrum: Zigzag banding of lights resembling walls of fortified medieval towns that marks the edge of scintillating scotoma.
        - Scintillating scotoma: a localized area of blindness edged by brilliantly colored shimmering lights (teichopsia), which gradually expands to encompass a larger portion of visual field.
    - Somatosensory aura:
      - Paresthesias (pins and needles or numbness) usually begin in hand and move up the arm and face over several minutes.
    - Dysphasic aura
      - Speech disturbance.
- Neurotologic Features (Vestibular migraine)\textsuperscript{76, 98, 111, 178, 202}
  - Vertigo is quite common in migraine. 26.5% of migraine patients have vertigo.
  - Migraine headache may be associated with episodic vertigo, motion sensitivity, and nonspecific dizziness such as swimming or rocking sensation inside the head.
  - Patients usually give h/o lifelong or childhood motion sensitivity. Swimming or rocking sensation provoked or aggravated by motion such as amusement park rides, back seat riding in a car, or reading in car, escalators; moving
visuals in supermarkets, shopping malls, passing trains. Due to motion sensitivity during the attack of headache patients prefer to lie still in a dark, calm and quite room.

- Episodic vertigo (rotational or to-and-fro) usually lasts for few minutes to several hours but in rare cases last for few seconds to few hours.
- Vertigo is associated with postural imbalance and unsteadiness with motion sensitivity or visual hallucinations
- Vertigo after taking certain food.
- Multiple attacks may be several per month.
- 62% had only vertigo, and 16% has vertigo with tinnitus and episodic hearing loss and ear fullness.
- Vertigo usually occurs during the headache (47%) but may occur during the headache free interval (36%) or preceding the headache (15%).
- The first attack of vertigo can occur between the ages of 7 and 72 years.
- 68% vestibular migraine patients give positive family h/o migraine.
  - Auditory symptoms:
    - Phonophobia, which may be during headache or headache-free interval, is most common. Hearing discomfort thresholds are significantly lowered.
    - Tinnitus is common (15% in migraine and 60% in basilar migraine).
    - Low frequency SNHL.
  - Some patients show central ocular motor signs between attacks.
  - Vertical and horizontal impaired pursuit
  - Gaze-evoked nystagmus
  - Moderate positional nystagmus
  - Spontaneous nystagmus with Frenzel glasses.
  - Caloric testing: Reduced response unilateral (18% to 60%) and bilateral (4% to 12%).
  - Rotational testing: Prolonged duration of response is the most common abnormality.
  - Migraine-precipitants before vertigo: food trigger, sleep irregularities, hormonal change.
  - Chronic nonspecific disequilibrium, unsteadiness, and sensitivity to motion of self or environment similar to psychophysiologic dizziness.
  - Gait and balance are normal though complaining of unsteadiness.

**DD**

Psychophysiologic dizziness.

**Migraine and Meniere’s Disease (MD)**

The diagnosis of both the diseases is based on clinical criteria and there is no definitive diagnostic test.
Lifetime prevalence of migraine was found to be significantly higher in patients with MD. Migraine headache occurred in during the vertigo attacks in 28% cases of MD. Vertigo attacks may be accompanied with headache, photophobia, or aura in 45% patients of MD. The presence of visual or somatosensory aura or headaches associated with episodic vertigo indicates migraine. Migraine has even been associated with sudden drop-attack falls attributed to the otolithic crises of Tumarkin in cases of MD. Fluctuating cochlear symptoms were seen in 12% to 38% cases of migraine. Baloh suggested a common pathophysiologic mechanism for the patients who have migraine with their MD. A defective ion channel with predominant expression in brain and labyrinth could lead to a local buildup of extracellular potassium that may result in the following:

- Spreading depression -- a wave of decreased cerebral perfusion that slowly passes across the cortex.
- Paroxysmal osmotic disequilibrium causes endolymphatic hydrops
- Toxic effects of increased perilymphatic potassium on inner ear hair cells.

i. Basilar Migraine

- Subtype of migraine with aura
- Recurrent headache usually occipital.
- This is associated with minimum two of the multiple neurologic symptoms of brain stem, cerebellum and occipital lobe (supplied by basilar artery).
- Consciousness is impaired quite often (77%)
- Symptoms of vertigo, fluctuating hearing loss and tinnitus may make basilar migraine difficult to distinguish from Meniere's disease and vertebrobasilar TIAs.
- Tinnitus is common (15% in migraine and 60% in basilar migraine).
- Low frequency hearing loss in 80%, bilateral (46%) or unilateral (34%). Hearing loss was fluctuating in 60%.
- Sudden profound unilateral SNHL occur occasionally. Migraine related vasospasm can, cause cochlear infarction.

ii. Vestibular Migraine

- Subset of basilar migraine

There is no agreed upon classification for vestibular migraine, which is under diagnosed. International Headache Society (IHS) classification does not specifically include vestibular migraine. Just coincidence of migraine and vertigo is not enough to label the case as vestibular migraine, as other diagnoses should be ruled out. In absence of episodes of vertigo, sensation of abnormal motion or imbalance is usually related to head movement. The proposed criteria for definite diagnosis of Vestibular migraine (Migrainous vertigo) are following:
1. Episodic vertigo of moderate (interfere with daily activities) or severe (prohibit daily activities) nature: rotational, illusory self or object motion, positional vertigo, head motion intolerance

2. Lifetime diagnosis of migraine by (IHS) criteria

3. Minimum one of the following migrainous symptoms during at least two vertiginous attacks
   a. Migrainous headache
   b. Photophobia
   c. Phonophobia
   d. Aura such as visual, somatosensory, or dysphasic

4. Other causes ruled out by necessary investigations

The proposed criteria for probable diagnosis of Vestibular migraine (Migrainous vertigo) are following:

1. Episodic vertigo of moderate (interfere with daily activities) or severe (prohibit daily activities) nature: rotational, illusory self or object motion, positional vertigo, head motion intolerance

2. Other causes ruled out by necessary investigations

3. At least one of the following
   i. Migraine according to (IHS) criteria
   ii. Migrainous symptom during vertigo
   iii. Migraine specific precipitants of vertigo such as food triggers, sleep irregularities, hormonal changes
   iv. Response to antimigraine drugs.

DD: Psychophysiological dizziness.

iii. Migraine Equivalents and Channelopathies (Episodic Ataxias)

Rare. Dominantly inherited channelopathies present with episodes of vertigo and ataxia.

1. **Episodic Ataxia type 1 (EA-1)**
   - Brief episodes of ataxia lasting minutes with interictal myokymia (muscle rippling in between the attack) in eyelids and fingers.
   - Missense mutations in human potassium channel gene (KCNA1 gene on chromosome 12q) involving brain. First reported mutation in human potassium channel gene and first ion channel mutation involving the brain.

2. **Episodic Ataxia type 2 (EA-2)**
   - Longer episodes of ataxia lasting hours with nystagmus (downbeat, gaze-evoked, or rebound) that persists between the spells (interictal nystagmus).
   - May be associated with vertigo, nausea and vomiting.
   - Episodes can be triggered by stress and exercise.
   - Relieved by long treatment with acetazolamide.
   - Some patients present with gradually progressive ataxia in later life.
- α1A subunit of CACNA1A voltage-gated P/Q calcium channel on chromosome 19p, which is expressed heavily in cerebellar Purkinje Cells.

**Familial Hemiplegic Migraine (FHM)**
- Most missense mutations in CACNA1A have been associated with FHM, a rare type of migraine with aura.
- FHM is characterized by recurrent episodes of migraine headache and hemiplegia. Minor head injury may trigger severe attacks, which may be associated with lethargy and coma.
- Some family members have only migraine with aura.

3. **Benign Paroxysmal Vertigo (BPV) of Childhood**
- BPV was described in 1964.
- Age of onset is usually 2 to 4 years.
- Frequency of attacks can be every few days to every few months. Episodes gradually decrease and disappears by 5 to 10 years.
- Recurrent sudden brief (<5 minutes) episodes of vertigo in children (2-4 years) without loss of consciousness or abnormal neurologic sign.
- May be associated with pallor, nausea or diaphoresis.
- Small children may not be able to describe vertigo but may grasp something nearby for support, sway or refuse to stand.
- Vertigo may be followed by sleep and then child becomes normal.
- Family H/O migraine is often (83%) present. Later on child may develop migraine headache, abdominal pain or cyclic vomiting.

4. **Benign Recurrent Vertigo (BRV)**
- Recurrent episodes of vertigo, nausea and vomiting (minutes to days) generally begin in adulthood often on awakening in the morning.
- Vertigo episodes may last from minutes to days.
- BRV affects women twice as often as men.
- Common around menstrual period.
- Attacks precipitated by common migraine triggers.
- H/O Migraine attacks in patient/family usually present.
- Familial BRV, a migraine syndrome is likely to be inherited in an autosomal-dominant fashion with decrease penetrance in men.

5. **Benign Paroxysmal Torticollis of Infancy (BPTI)**
Migraine equivalent having cervical dystonia.
- Recurrent episodes of head tilt with vomiting, pallor, ataxia lasting hours to days.
- Begins in first year of life but resolves by 5 years of age.
- After the 5 years of age, child may develop BPV and later on migraine headache.
d. Frontal Gait Disorder of Elderly or Gait Apraxia or Lower-half Parkinsonism - Multiple Lacunar Infarcts

Higher level gait disorders

Frontal Gait Disorder or Frontal disequilibrium
Gait of frontal lobe lesions is common in following disorders

- Normal pressure hydrocephalus
- Multiple lacunar infarcts
- Subcortical small vessel disease

Lower-half Parkinsonism or Gait apraxia
Difficulty in initiating movement, wide based gait with an upright posture of trunk, short shuffling steps with normal or exaggerated arm swing.

2. Inflammatory

a. Multiple Sclerosis (MS) 107, 110, 134, 297

Demyelinating disorder of CNS, where myelin is formed from oligodendroglia. Schwann cells are unaffected. Demyelinating plaque affects vestibular nuclei, cerebellum and its peduncles, and cranial nerves.

Etiology
Cause not known but autoimmunity, infection and heredity may play a role.
Age: 15 to 50 years. Peak at age 24.
Sex: Females are more affected than males (2:1)
Predilection for northern European heritage

Clinical

- Usually manifests as either relapsing and remitting or progressive.
- CNS dysfunctions are disseminated in time and space.
- Myriad neurological features include
  - Pyramidal signs: Weakness, hyperreflexia, and Babinski sign
  - Sensory tracts: Loss of vibration or joint position sense.
  - Cerebellum: Ataxia and intention tremor
  - Brainstem: Cranial nerves involvement.
  - Partial transverse myelitis: Paraparesis
  - Common findings: Blindness and diplopia
    - Optic neuritis: Monocular vision loss
    - Bilateral internuclear ophthalmoplegia: Diplopia
- Vertigo occurs in 50% cases but is initial symptom in only 5% cases.
  - Sustained over days to weeks
  - Paroxysmal or positional: Patient may have positional nystagmus, some times with vertigo. Positional nystagmus with vertigo may be the first manifestation of MS176. Confirm the diagnosis through the observation of classic eye movements in association with Dix-Hallpike maneuver.
  - Prolonged spontaneous attacks of vertigo
  - Involvement of intrapontine portion of vestibular nerve or nucleus may resemble vestibular neuritis: Vertigo (hours to days), vomiting, imbalance, direction-fixed (toward normal side) horizontal-torsional nystagmus, and canal paresis (caloric test).
The clinical manifestation of selective involvement of vestibular nuclei may be indistinguishable from acute peripheral vestibular disease.

Distinguishing feature: This central nystagmus may not be suppressed by visual fixation.

• Demyelinating plaque involving vestibular nuclei, cerebellum and its peduncles: Severe ataxia, direction-changing nystagmus, intention tremor or pyramidal signs.

• Paroxysms of vertigo are sometimes precipitated by hyperventilation and are managed with membrane stabilizing drugs such as carbamazepine or phenytoin, or with acetazolamide.

• DD: The inflammatory disorders of labyrinth, which can present with recurrent vertigo, include syphilis, Cogan’s syndrome, and sarcoidosis and Susac’s syndrome (retinocochleocerebral vasculopathy). The connective tissue lesions, which can have recurrent vertigo, include SLE, RA, Behcet’s disease, and antiphospholipid syndrome.

• Sudden hearing loss may accompany vertigo.

• Eyes
  - Pendular nystagmus, which often causes oscillopsia and poor vision, is a common feature.
  - Depending on the horizontal and vertical components, eye movements may be oblique, elliptical, or circular.

• Patients usually present with weakness, ataxia, vision loss in either eye, diplopia (double vision), or sudden hearing loss.

Supportive Investigations:

• There is no diagnostic test.
• MRI: Demyelinating plaques
• CSF: Oligoclonal bands or a raised IgG index
• Evoked potential: Slowing along visual, auditory, or somatosensory pathways due to demyelination.

b. Superficial Siderosis

Rare disease
Accumulation of iron, in cases of recurrent subarachnoid bleeding from occult vascular malformation, damages vestibulocochlear nerve and cerebellar cortex.
Patient’s characteristics include progressive bilateral SNHL and ataxia.
MRI: Hemosiderin deposition over the cerebellar hemispheres.

3. Infectious

a. Intracranial Complications of Otitis Media
b. Febrile Illness – Cerebellar Ataxia
c. Acute Viral Syndromes
d. Meningitis
e. Cerebellar Abscess (Fungal or Bacterial)
Coarse horizontal nystagmus, dysmetria, dysdiadokokinesia, or action tremor.

f. Tabes Dorsalis: Rapid bilateral hearing loss with wide-based gait

4. Neoplastic (Brain tumors)  
   o Brain tumors though most feared are rare causes of dizziness.
   o Usually patients present with progressive focal neurologic deficit through weeks to months.
   o Because of hemorrhage in tumor, some patients present with sudden stroke like neurologic deficit.
   o MRI with gadolinium can detect virtually all brain tumors.
   o Cerebellopontine angle and temporal bone tumors are included in peripheral vestibular disorders.
   o Neuropathy or degenerative joint disease can also cause imbalance.

a. Cerebellar Tumors  
   - Main types: Astrocytomas, ependymomas, medulloblastomas, hemangioblastomas, and metastases.
   - Most primary cerebellar tumors occur in children.
   - Metastatic cerebellar tumors are common in adults.

   Clinical
   - Occipital headache radiating forward is the most common presentation.
   - Neck stiffness and limitation of range of motion suggest incipient tonsillar herniation.
   - Ipsilateral appendicular ataxia (hemispheric involvement), truncal and gait ataxia (midline vermis tumors).
   - Positional vertigo and nonfatigable downbeat nystagmus may occur.
   - Cyclic vomiting or vomiting on lying down may occur.
   - Secondary invasion or pressure on brainstem can cause lower cranial nerve palsies (dysarthria and dysphagia) or pyramidal tract findings.
   - Acute obstructive hydrocephalus due to pressure obstruction of fourth ventricle results in rapid neurological deterioration.

b. Paraneoplastic syndromes (Paraneoplastic encephalomyelitis)  
   Autoimmune reaction occurs probably against antigens coexpressed by tumor cells and neurons. Primary malignancy must be sought actively in cases of subacute cerebellar syndrome.

Paraneoplastic opsoclonus-myoclonus (POM)
Brief jerky involuntary movement of limb and eyes

i. Paraneoplastic Cerebellar Degeneration (PCD)

   Clinical
   - Cerebellar-type ocular motor and vestibular abnormalities, dysarthria, and severe appendicular and gait ataxia.
   - Begin abruptly and progress rapidly through several months.
   - Patients may not be able to walk or feed themselves.

   Diagnosis
   Development of anti-Purkinje cell antibodies (PCAs) in following asymptomatic or undiagnosed malignancies:
   2. Carcinoma
      a. Ovary – seropositive for PCA-1 (anti-Yo) antibodies
b. Breast - seropositive for PCA-1 (anti-Yo) antibodies

c. Lung – small-cell cancer – PCA-2 and ANNA-1 (anti-Yo) antibodies

ii. Paraneoplastic Opsoclonus - Myoclonus (POM)

**Etiology**
1. Children with Neuroblastoma
2. Women with breast and gynecologic cancer
3. Immunology related to preceding infection might play a role when no occult malignancy is found

**Clinical**
- Opsoclonus (saccadomania), which occurs due to the dysfunction of omni pause cells (inhibitory saccade-related neurons) within brainstem, refers to back-to-back usually of large amplitude multidirectional conjugate saccades.
- Myoclonus refers to jerky involuntary limb movements.
- Patients usually have vertigo before the appearance of opsoclonus and ataxia.

5. Craniocervical Junction Disorders

**Chiari Malformation**
- Group of hindbrain disorders associated with abnormalities of craniocervical junction, which present with acquired ataxia.
- In Chiari I malformation, which is mildest and most common type, there occurs displacement of deformed cerebellar tonsils more than 5 mm caudally through foramen magnum

**Clinical**
- Patients often have no symptoms and diagnosis is incidental on MRI.
- Patients may have constant or slowly progressive nonspecific dizziness and gait instability that worsen with neck extension.
- Vertigo, tinnitus, and hearing loss are uncommon.
- Patients often have dizziness, vertigo, neck pain, or headaches brought on by Valsalva maneuvers.
- Gag reflex may be absent.
- Typical oculomotor findings localizing to the vestibulocerebellum (tonsils or paraflocculi).
- Downbeat nystagmus with head hanging or neck extension and spontaneous central vestibular nystagmus may occur in some cases.
- Midline sagittal T1W MRI scan diagnostic: Cerebellar tonsils below foramen magnum.

6. Inherited (Hereditary) Ataxias

Family history is usually present though spontaneous mutations and variable penetrance without a family history are also reported.
Ataxia is present in many inborn metabolic and mitochondrial disorders.

a. Autosomal Recessive
i. Friedreich Ataxia

**Etiology**
- GAA repeat expansion in frataxin gene.
Clinical
Clinical features, which include reduced VOR gain, saccadic intrusions (square-wave jerk) axonal sensory neuropathy with areflexia, dysarthria, Babinski sign, diabetes and cardiomyopathy, usually appear before 20 years of age but in some cases as late as 6th decade.

b. Adult Onset Autosomal Dominant or Spinocerebellar Ataxias (SCAs)
Spinocerebellar ataxias (SCAs) are classified by sequential numbers SCA1 through SCA22, which are assigned as a new genetic locus is reported. Some SCAs have distinguishing features such as following:
- SCA2 – very slow saccades
- SCA3 – dystonia or Parkinsonism
- SCA6 – pure cerebellar syndrome

Given the significant phenotypic overlap, genetic testing is warranted and is available for SCAs 1, 2, 3, 6, 7, 8, 10, and 17.
SCAs can be further categorized into three discrete groups based on pathogenesis.

i. Polyglutamine disorders
SCAs 1, 2, 3, 7, and 17 result from toxic stretches of polyglutamine.

ii. Channelopathies (calcium or potassium)
SCA6 and episodic ataxia types 1 and 2.

iii. Gene expression disorders
SCAs 8, 10, and 12 result from the repeat expansions outside of coding region.

7. Metabolic
a. Wernicke’s Encephalopathy

Etiology
- This neurologic emergency (combination of midline cerebellar and either proprioceptive or vestibular impairment) is caused by thiamin deficiency due to poor oral intake.
- Common in chronic alcoholic, undergoing chemotherapy, anorexia, hyperemesis gravidarum or intravenous glucose load.

Pathology
Neuronal loss and gliosis in the nuclei of CN III, IV, VI, and VIII (earliest in lateral vestibular nuclei), cerebellum, and vestibulospinal tracts.

Clinical
- Characterized by triad of ophthalmoplegia, confusion and dramatic gait ataxia, which increases with eye closure or darkness.
- Finger-to-nose and heel-to-shin tests are relatively normal.
- Patient is unable to stand or walk without support.
- Vertigo uncommon
- Ocular motor findings: Weakness of abduction, gaze-evoked nystagmus, internuclear ophthalmoplegia, upbeat or downbeat nystagmus often changing with convergence.
- Horizontal and vertical gaze palsies may develop to ophthalmoplegia.
- Caloric, rotational and head thrust tests: Vestibular loss.
Diagnosis
- Clinical
- MRI: Fluid-attenuated inversion recovery (FLAIR) and T2W images
  o High signal intensities in medial thalami, mamillary bodies, tectum of mid brain, vestibular nuclei, and periaqueductal region.
  o Atrophy of superior cerebellar vermis in chronic alcoholics
b. Diabetes
c. Vitamin B-12 Deficiency
d. Vitamin E Malabsorption
e. Celiac Disease
f. Hypothyroidism
g. Hypoparathyroidism
h. Hypoglycemia
i. Hyperventilation
8. Toxic
   a. Medications
      i. Methotrexate: Disequilibrium - Brainstem and cerebellar toxicity
      ii. Amiodarone: Antiarrhythmic drug that can cause disequilibrium and other neurological symptoms.
      iii. Anticoagulants: Vertigo - hemorrhage into inner ear or brain
      iv. Antiepileptics: Carbamazepine (Mazetol), phenytoin (Dilantin), primidone. Disequilibrium due to cerebellar toxicity.
      v. Antihypertensives and Diuretics: Causes near fainting. Reduced cerebral blood flow occurs due to postural (orthostatic) hypotension.
      vi. Tranquilizers and Antihistamines: Causes intoxication due to central nervous system depression.
         1. Tranquilizers
            a. Barbiturates
         2. Antihistamines
            a. Tricyclic amines
b. Alcohol
   • Disequilibrium due to Cerebellar toxicity: Chronic alcoholism can cause not only cerebellar degeneration but also predispose the patient for Wernicke’s encephalopathy. The relatively selective toxicity to anterior cerebellar vermis predominantly leads to gait ataxia with relative sparing of limb coordination.
   • Vertigo due to change in cupula-specific gravity
   • Intoxication (Central nervous system depression): It can produce horizontal positional nystagmus by altering the specific gravity of the cupula relative to surrounding endolymph.
9. Degenerative
   a. Parkinson’s Disease (PD)
      Pathology
      Degeneration of basal ganglia and substantia nigra and Lewy body cytoplasmic inclusions.
      Types
      • Idiopathic
- Parkinsonism-plus disorders
- Toxic
- Metabolic
- Vascular

**Clinical**

i. Characterized by tremors at rest (Pill-rolling movements of fingers), rigidity, bradykinesia (slowness of movement), and postural instability

ii. Stooped posture, difficulty initiating gait, festinating gait, shuffling, reduced arm swing and difficulty in turning are common.

iii. Patient may fall forward or backward. Nudging also may cause fall.

iv. Ocular motor abnormalities: Square wave jerks, hypometric saccades, and impaired smooth pursuit.

**b. Progressive Supranuclear Palsy**

**Age of onset:** 6th or 7th decade

**Pathology**

Neurofibrillary tangles with neuronal loss and gliosis in areas of subcortical and brainstem regions and tau-positive neuronal and glial inclusions.

**Clinical**

i. Unsteady gait with propensity to sudden falls (esp. backward), dysarthria (slow, slurred speech with a strained voice), cognitive and emotional changes and visual complaints are most common.

ii. Akinesia, axial rigidity, and brisk facial reflexes, apraxia of eyelid opening, lid lag, blepharospasm, and inability to suppress blinking of eyes on exposure to bright light are also common findings.

iii. A wide-eyed stare of astonishment with elevated eyebrows, and forehead furrows and typical dysphonia are suggestive of diagnosis.

iv. Midbrain vertical gaze centers involvement gives rise to vertical supranuclear gaze palsy.

v. Following ocular motor findings may be present: Slowing of vertical saccade particularly downward; slow and hypometric horizontal saccades; normal VOR; convergence absent; impaired vertical smooth pursuit.

**c. Multiple Systems Atrophy (MSA)**

**Pathology**

Neuronal multisystem degeneration and glial cytoplasmic alpha-synuclein inclusions.

**Clinical**

MSA is characterized by progressive autonomic deficits, especially orthostatic hypotension, impaired GIT motility, urinary and sexual dysfunction

**Types**

i. MSA-P: Striatonigral degeneration results in features of Parkinsonism. Rest tremors are absent and condition does not respond to levodopa.

ii. MSA-C or Olivopontocerebellar Atrophy (OPCA): Imbalance, falls, ataxia and orthostatic hypotension may occur. Positional downbeat nystagmus common.
iii. Shy-Drager
iv. Normal Pressure Hydrocephalus
d. Degeneration of anterior cerebellar vermis
Chronic alcoholics
Gait ataxia predominates with relative sparing of limb coordination.

10. Epilepsy
Partial Seizures - Petit Mal - Tornado Epilepsy
A rare cause of recurrent vertigo.
May occur without other symptoms or altered consciousness.
Dizziness and vertigo are common side effects of anticonvulsants and other drugs, which are advised for epilepsy.

DD
Basilar Migraine

11. Trauma
Post-concussion Syndrome
It is more frequent after minor head trauma and presents with
- Headaches,
- Irritability,
- Depression,
- Lassitude, and
- Vertigo

12. Physiologic Dizziness
Mismatch among the sensory signals (physiologic stimulation of vestibular, visual, and somatosensory systems) in normal persons results in disorientation, imbalance and vegetative symptoms.
a. Motion Sickness
- This common form of physiological dizziness occurs in susceptible individuals usually with prolonged vestibular stimulation. It may also be caused by visual stimulation.
- Complete bilateral vestibular loss makes the patient resistant to motion sickness.

Migraine and Motion Sickness
- Migraine patients are more prone to motion sickness.
- Motion sickness in children may be the starting feature of migraine.

Clinical
- Dizziness, fatigue, pallor, cold sweats, salivation, nausea and vomiting develop aboard a ship, in a car, on an airplane, or in space.

Aggravating factors (visual vestibular conflict)
- Visual vestibular conflict occurs when person is not able to visualize movement viz. sitting in an enclosed cabin of a ship.
- Reading in a car and riding in the back seat of a car.

Relieving factors (minimizing visual vestibular mismatch)
- Standing on deck of the ship and focusing on the horizon or land.
- Sitting in the front seat of a car and looking off in the distance.
- Minimizing head movements by resting the head against the headrest of a vehicle.

b. Mal de Debarquement (MDD) Syndrome
- Sickness of disembarkment presumed to be caused by multisensorimotor adaptation and habituation to a new or abnormal motion environment.
- Inappropriate sensations of movement (rocking, swaying, disequilibrium or unsteadiness) for a month (may last for years) after sea voyage, extended train, air or car travel.
- Patients feel better in moving vehicles and while walking.
- Usually affects women. Mean age of onset is 5th decade.

13. Psychophysiologic and Psychogenic Dizziness

Vestibular symptoms can be the cause as well as manifestation of psychiatric diseases\textsuperscript{317}.

a. Psychophysiologic Dizziness\textsuperscript{93}

There occur abnormal integration of sensory information, which is used for balance and orientation.
Patients may have sensations of floating, rocking, or swimming, spinning vertigo, giddiness, imbalance, and being removed from body.
These sensations may be either continuous or intermittent and may worsen with stress or fatigue.

Aggravating factors
- Walking on bright or patterned floor or supermarket aisle.
- Driving on busy roads
- Seeing too-much motion films or TV programs.
- Shopping in crowded places
- Transient or sustained vestibular disorders such as vestibular neuritis.
- Symptoms are associated with anxiety and behavioral changes.

Phobic Postural Vertigo (PPV)\textsuperscript{59}

PPV patients are obsessively preoccupied with their psychophysiologic dizziness predominantly subjective postural imbalance without falls.

Clinical

The following are the characteristic features:
- Subjective imbalance while standing or walking. Balance tests normal.
- Episodes of fluctuating unsteadiness lasting seconds to minutes or momentary perceptions of illusory body perturbations.
- Though can be spontaneous, following perceptual provoking factors (develop rapid conditioning, generalization and avoidance behavior) are often present:
  - Places: Bridge, staircase, empty room, or street
  - Social situations: Crowd, shopping malls, concert or restaurant.
- Distressing vegetative symptoms develops with or without anxiety.
- Personality: Obsessive-compulsive type, labile affect, and mild depression are common.
- Begins after particular emotional stress or serious illness or organic, vestibular lesion.

DD

Panic disorder or agoraphobia.

b. Psychogenic Dizziness\textsuperscript{93}
- Experience is required to diagnose psychogenic dizziness as anxiety and depression can be not only a reaction but also the cause of dizziness.
- Rotational vertigo with direction specific fall, nausea and vomiting are uncommon.
- Dizziness occurring only in combination with cluster of recognized psychiatric symptoms (anxiety or panic disorder), which are not related to vestibular dysfunction (such as BPPV or vestibular migraine) favor the diagnosis of psychogenic dizziness.

Following clinical characteristic should raise the suspicion of psychogenic vertigo:
- Certain stimuli or social functions trigger episode of dizziness.
- Rotational vertigo without spontaneous nystagmus
- Dissociation between subjective and objective disequilibrium
- Excessive anxiety or fear of impending death

Hyperventilation

Though hyperventilation usually induces dizziness in anxious or phobic individuals it can also cause dizziness in peripheral or central vestibular disorders.

Rapid drop in Pco₂ is presumed to result in cerebral vasoconstriction.
  - Common cause of dizziness in anxious young people
  - Symptoms include giddiness, lightheadedness, feelings of suffocation, perioral and acral (extremities such as limbs, fingers, or ears) paresthesias (sensation of burning, pricking, tickling, numbness or tingling).
  - Voluntary hyperventilation can reproduce symptoms but may also provoke symptoms from peripheral or central vestibular disorders.

C. Anxiety disorders

- An anxiety is very obvious in cases of panic attacks without any cue or significant avoidance behavior.
- Among all the anxiety disorders, panic disorder is most commonly associated with vertigo.

Generalized Anxiety Disorder

- These patients usually when sitting alone or thinking over their problem, develop vague sensations of floating or giddiness along with fatigue.
- Dizziness, which lasts for years, may vary from time to time.

Panic Disorder

The diagnosis of panic disorder requires two or more panic attacks, which have following characteristic features:
- Onset: Abrupt with fear of dying or losing control and usually spontaneous without any cue. Situational trigger (cue) is present when a phobic patient is exposed to feared situation.
- Autonomic symptoms: Palpitation, sweating, nausea
- Hyperventilation symptoms: Dizziness, shortness of breath, chest pressure, faintness, depersonalization (losing his own identity or own reality in relation to others), perioral and acral (extremities such as limbs, fingers, or ears) paresthesias (sensation of burning, pricking, tickling, numbness or tingling).
- Subsequent worry about future attacks or change in behavior.

**Agoraphobia**
(G. agora, marketplace, + phobos, fear)
- Irrational fear of leaving the familiar setting of home, or venturing into the open; often associated with panic attacks.
- Patient of this type of panic disorder avoids or endures with great distress the feared situation.

d. **Depressive Disorders**
- Depression, which is not uncommon in patients of chronic vestibular disorders, can cause psychogenic dizziness.
- Because of the suicidal tendencies, these patients are referred for psychiatric treatment.

14. **Presyncope Dizziness (Global Cerebral Hypoperfusion)**
- Reduction in blood flow to entire brain leads to faintness or presyncope (before fainting or losing consciousness), which clinically present as lightheaded sensation that may be accompanied with giddiness, generalized weakness, and pallor.
- The common causes of presyncope include cardiac, orthostatic hypotension, vasodepressor effect, or hyperventilation. Uncommon causes include focal carotid or vertebrobasilar stenosis or occlusion.
- In contrast to vertigo there is no illusion of motion.
- Dizziness may be spontaneous, positional or have triggers.

a. **Vasovagal Presyncope or Vasodepressor (Vasovagal) Syncope**
- Most common cause of syncope
- Symptoms: Nausea and sweating followed by lightheadedness, dimming of vision, and pallor
- Symptoms are usually situational and provoked by emotional stress, hunger, heat, acute pain, or prolonged standing.
- Frank syncopal attack is usually avoided by immediate lying down.

b. **Reduced Cardiac Output**
- Provoked by Exertion and associated with palpitation or chest pain
- Causes may include
  - Cardiac arrhythmias
  - Outflow obstruction
  - Constrictive pericarditis
  - Cardiomyopathy

c. **Orthostatic Hypotension**
- Inadequate autonomic factors fail to maintain blood pressure (Fall of BP) and cerebral perfusion when person is standing upright.
- Patients who demonstrated a fall in diastolic or systolic blood pressure of over 10 mmHg after standing from a supine posture were diagnosed as postural hypotension.

Causes
Autonomic nervous system dysfunctions in following conditions:
1. Multiple system atrophy (MSA)
2. Diabetes mellitus
3. Amyloidosis

Medications:
1. Diuretics
2. Antihypertensives
3. Levodopa.

Hypovolemia from dehydration or blood loss
d. Hyperventilation
   Discussed under psychogenic dizziness
e. Hypoglycemia
   - Fasting especially in diabetics
   - Symptoms of lightheadedness, lethargy, confusion, and shakiness
   - Usually caused by insulin or sulfonylurea in diabetics.

15. Multisensory Disturbances
Multisensory disequilibrium is the cumulative effect though mild of multiple sensory deficits, which include following:
a. Peripheral Neuropathy: impaired proprioception
b. Lumbar root disease
c. Posterior column disease: Romberg sign is present in patients who are dependent on vision for the maintenance of balance.
d. Retinopathy and cataracts: Impaired vision
e. Reduced vestibular responses in elderly.
Other factors include
f. Cervical spondylosis: Myelopathy, weakness and sensory deficits.
g. Cervical or Thoracic Myelopathy
h. Aging: Cerebellar atrophy and diffuse small vessel ischemia

Nonneurologic causes
i. Degenerative joint disease

Dynamic posturography: To know how much patient depends on each sensory subsystem for balance.

16. Ocular Motor Disorder
a. Superior Oblique Myokymia (SOM)
   This nonvestibular syndrome, which is not central in origin, presents with brief spells of oscillopsia or diplopia.

Etiology
Cause may be some neurovascular compression similar to trigeminal neuralgia and hemifacial spasm.

Clinical
- Paroxysmal monocular oscillopsia and microtremor for <10 seconds may occur many times a day.
- Intermittent contraction of superior oblique muscle results in monocular torsional and vertical eye movements.
- Patients usually present with brief (<10 seconds) recurrent (even many times per day) episodes of monocular blurring of vision or tremulous sensations in one eye.

Diagnosis
MRI: Compression of trochlear nerve at its exit by a branch of superior cerebellar artery in some cases.

17. Miscellaneous
a. Musculoskeletal Vertigo (Cervical Vertigo)
Patients with neck pain or occipital headache but no features of otological or neurological disease in whom vertigo is provoked by changes in neck position are diagnosed musculoskeletal vertigo. Cervical spondylosis is common in old people and too often blamed for symptoms. It should be considered only when vertigo is clearly associated with movements of the neck (not the head) and appreciable radiological changes.