NEUROLOGY

C O N T E N T S

I. INTRODUCTION

This section is written for physicians who treat patients with neurological complaints. It covers both common neurological complaints and life threatening neurological disorders, and provides basic guidelines for adequate diagnosis and treatment of these conditions. As neurological consultation and expensive neurodiagnostic testing are often unavailable, reliance on history and examination will be emphasized.

General Neurological Evaluation

The primary goals in neurological evaluation are lesion placement and differential diagnosis determination. Place the lesion in one of the following five areas:
- The cerebral hemispheres (i.e., supratentorial)
- The brain stem or cerebellum (i.e., infratentorial)
- The spinal cord level
- Peripheral nerve and/or nerve root Muscle

Be aware that isolated musculoskeletal problems may present as neurological complaints.

A convenient mnemonic for remembering the neurological differential diagnosis is VIN DIITTCH MD:

V   Vascular
I   Infectious
N   Neoplastic
D   Demyelinating
I   Idiopathic
I   Immune
T   Trauma

T Toxic
C Congenital
H Hereditary
M Metabolic
D Degenerative

Obtain a thorough history, with specific emphasis on the chief complaint. If possible, establish the time course, including onset, progression, duration, recurrence, and resolution. The time course is often a clue to the most likely etiology. For example, a chronic, slowly progressive condition may indicate a neoplastic or degenerative process, whereas an intermittent deficit may suggest a vascular or demyelinating condition. With severe, sudden, or recurrent neurological complaints, consider potentially life threatening conditions.

The mnemonic LEARNIT is useful when taking a neurological history, particularly for pain syndromes:

L Location
E Exacerbating factors
A Alleviating factors
R Radiation
N Nature
I Intensity
T Timing

In the past medical history, explore potential occupational hazards such as toxic substances and solvents, high intensity noise, and travel to areas endemic for tropical diseases.

Include a general physical examination with attention directed to the head, spine, and extremities. Consider congenital and hereditary conditions in patients with dysmorphic facial features, subtle differences in extremity size, flat or highly arched feet, or other similar abnormalities. The neurological examination begins with the mental status examination, which may only consist of the overall response of the patient to the examiner. However, if the patient complains of specific problems of thinking, such as memory loss or decline in work performance, examine mental status further. Formal mental status tests include the Mini Mental Status Exam, the Galveston Orientation and Amnesia Test (GOAT), and Halstead Reitan Test Battery.

Mental Status Examination
The mental status examination includes level of alertness, affect, physical appearance, and orientation to person, place and time. Evaluate memory function with immediate, short term, and remote memory tests. Test immediate recall with forward and reverse "digit span", by having the patient recite a short list of numbers forwards and backwards. Short term memory is tested with recall of a list of objects or a short story after three minutes. Test remote memory by asking about past presidents or specifics from the member's service or health records. Ask the patient to

interpret proverbs or make comparisons between similar objects to test judgment, insight, and abstracting ability. Test calculations with coin exchange problems such as the number of nickles in $1.35, or by having the patient repeatedly subtract seven from each successive answer, starting with 100.

Next, evaluate the cranial nerves. If the patient complains of visual difficulties, test visual acuity, monocular color vision, and visual fields. Compensate for refractive error with a pin hole or by re-refracting the patient. Measure pupil size in both light and dark backgrounds. Record pupil response both to direct light and to accommodation. The pupils should react the same with the same light source. Also ask if the light has the same brightness in both eyes. An unequal pupillary response or a subjective difference in light intensity is suggestive of an afferent pupillary defect (APD) or a Marcus-Gunn pupil. Cover each eye to detect a shade difference of a red-colored object, which indicates a subtle afferent pupillary defect. Test visual fields by finger count confrontation. To perform this test, place a number of stationary fingers in the central 30 degrees of vision, and ask the patient to count the total. Test all four quadrants separately in each eye. During the fundoscopic examination, examine the optic disk, macula, blood vessels, nerve fiber layer, and as much as the surrounding retina as possible. Also look for jerky movements of the fundus, which may represent nystagmus. Note that several conditions of disc elevation simulate papilledema, including myelinated nerve fiber, optic nerve drusen, and a hypoplastic disc.
Comment on eyelid symmetry during the extraocular muscle examination. Test the eye muscles in the six cardinal fields of gaze to through the full range of motion. Subtle clues of eye muscle imbalance include asymmetrical corneal light reflex and different scleral margins in the cardinal fields of gaze. Test the cardinal fields by turning the head with the eyes fixed. If available, formal testing with a phoropter, red lens, or Maddox rod with prism bars gives an even more accurate measurement of misalignment. Eye muscle imbalance may also be detected using the cover/uncover (tropia) or alternate cover (phoria) methods. Have the patient stare at the smallest letter visible on the eye chart, and cover and uncover the eye. Note eye movement from covered to uncovered. An eye that moves from inward to outward exhibits esotropia. Again, the eyes are tested in the six cardinal fields by turning the head with the eyes fixed on the eye chart. To perform alternate cover testing, occlude one eye and note latent deviation. Deviation from inward to outward is esophoria.

Test sensory Trigeminal (V) nerve by eliciting the corneal or sternutatory reflexes. The corneal reflex is tested by applying a wisp of cotton to the cornea, or by gently blowing on each eye separately. Get good separation with the edge of your open hand placed vertically on the paitent's nose, so you only blow on one eye. Test sternutatory reflex by sticking a small object into the nose and looking for a blink or cough. Test trigeminal motor function by observing the muscles of mastication including masseters, temporalis, and pterygoids, which move the jaw front, back, and side to side. Facial nerve testing includes tests of lacrimation (Schirmer Test), stapedial reflex (tested on audiograms), taste (anterior 2/3 of the tongue), and facial expression (wrinkle forehead, close eyes, smile, and purse lips). Test the Glossopharyngeal (IX) and Vagus (X) nerves by eliciting the gag reflex, by assessing the position of the palate at rest, by saying "AH", and by testing phonation (saying the consonants ba, da, fa, la, ga). Test the Spinal Accessory (XI) nerve

by having the patient turn his head to either side against resistance, and by shrugging his shoulders (trapezius muscles). Test Hypoglossal (XII) nerve function by having the patient protrude his tongue forward and to either side. Feel the force of the side-to-side protrusion directly through his cheeks as he bulges out his cheek with his tongue.

Cerebellar Station and Gait Testing
Cerebellar testing includes finger to nose, heel to shin, rapid alternating movements, and rebound (ability to hold extremity against changing loads). Gait testing probes cerebellar, motor, and sensory function. Normal gait is tested by having the patient walk and perform rapid turns. Test stress gait by having the patient walk on the outsides and insides of his feet, and perform duck walking. Stress gaits may elicit reduced arm swing or hand posturing which indicates subtle paresis. Be sure the patient is not distracted by loose gowns or pants. Attempts to hold up loose clothing can mimic posturing. Tandem gait testing, which examines axial cerebellar function, is performed by having the patient walk heel to toe like a tightrope walker, first with eyes open, then closed. Test station by having the patient stand with feet together (Romberg position) with eyes both open and closed. If station is done without difficulty, next test in the tandem position. Have the patient place one foot in front of the other with the eyes open, then closed (Tandem Romberg). Finally, test in the sharpened Romberg position, with one foot in front of the other, head tilted back toward the ceiling, and eyes first opened, then closed.

Motor Examination
The motor examination is designed to detect muscle weakness in a pattern which localizes the level of involvement to the central nervous system, spinal cord, peripheral nerve, or muscle. The motor examination begins proximally and proceeds distally. Start with neck flexion, extension, and rotation. Then test shoulder abduction, adduction, internal and external rotation, and shrugging. Test elbow flexion and extension, both in pronation and supination. Then examine flexion and extension of the wrist, followed by finger flexion, extension, and spreading. Move to the lower extremities. Test hip flexion and extension, abduction and adduction, followed by knee flexion and extension, ankle dorsi-flexion, plantar-flexion, and finally toe flexion and extension. Grade motor strength according to a 0-5 point scale:

0   no movement
1   a flicker of movement
2   movement of the muscle with gravity removed (ie parallel to the floor)
3   movement overcomes gravity but not resistance
4   movement against resistance
5   normal strength

Examine the muscles for tone, noting any stiffness, elasticity, rigidity, or cogwheeling. Also note any postural tremor, resting fasciculation, or atrophy.

Sensory Testing
The sensory system is divided into fine sensation, carried in the posterior columns of the spinal

cord, and course sensation, carried in the spinothalamic tract. Fine sensation includes vibration, proprioception, and two point discrimination. Cortical sensation, processed from signals from the fine sensory system, is tested by having the patient identify numbers written on the palms and soles (graphesthesia), or by identifying objects such as coins placed in the palms. Another test of cortical sensation is double simultaneous stimulation. Apply a light touch to the palm, first on one side, then on both sides simultaneously. Crude sensory function, carried in the spinothalamic tracts, is tested by light touch, temperature, and pin prick. Use the tuning fork as a cold object. Twist and break a tongue blade or cotton tipped applicator, or use an open safety pin (from a triangular bandage) to test sharp and dull. Discard after use. Avoid the Wartenberg wheel, unless you sterilize it after each use.

Reflex Testing
Reflex testing includes deep tendon reflexes, frontal release reflexes, and cutaneous reflexes. Frontal release reflexes include the glabellar sign, the snout or root reflex, the palmomental sign, and the Wartenberg reflex. Elicit the glabellar by tapping the forehead and observing persistent blinking. Test the root reflex by tapping on the lips or by scratching the corner of the mouth and looking for a "rooting" contraction. Elicit the palmomental sign by scratching the palm. Look for twitching of the mentalis muscle, just beneath the lower lip. Test the Wartenberg reflex by having the patient very gently flex his fingers against resistance. Crossing of the thumb toward the palm is positive.

Deep tendon reflex assessment of the upper extremities includes at least biceps tendon and triceps tendon testing. Other reflexes to test are the brachioradialis, elicited by tapping over the radial aspect of the forearm, and the deltoid and pectoral reflexes, tested by tapping over the deltoid and pectoralis muscles, respectively. Finger flexion reflexes can be seen with normal brisk reflexes. When testing finger flexion, include the Hoffman and Tromner signs. The Hoffman sign is triggered by flicking the middle finger away from the palm and observing a pinching movement between the thumb and index finger. Elicit the Tromner sign by flexing the middle finger separately from the rest of the fingers and flicking it toward the palm, again looking for a pinching movement between thumb and index finger. These two signs do not necessarily indicate pathology, but rather brisk muscle stretch reflexes. Asymmetry may be significant.

Deep tendon reflexes in the lower extremity include the quadriceps reflex (knee jerk) and the gastrocnemius reflex (ankle jerk). In addition, reflexes of the hamstring muscles (biceps femoris) can be tested. The plantar response, or Babinski sign, should also be tested. The Babinski sign refers to initial dorsiflexion of the great toe with spreading of the other toes. It is indicative of corticospinal tract dysfunction. Elicit the Babinski sign with a gentle stimulus to the lateral aspect of the sole, starting at the heel and moving upwards to the base of the little toe. The same reflex can be elicited by stimulating the side of the foot in a similar manner. This maneuver is called Chaddock's sign. Other techniques to elicit the Babinski sign include a brisk lateral abduction and release of the little toe, or a rapid, flicking motion applied to the third or fourth toe. As before, great toe dorsiflexion is abnormal or positive. These maneuvers are helpful if the patient's leg is

casted and you are unable to scratch the sole of the foot.

Test superficial cutaneous abdominal reflexes by scratching from the margins of each abdominal quadrant toward the umbilicus. Observe any quivering motion. The deep abdominal reflex is elicited by tapping over the anterior rectus abdominis muscle sheath and observing contraction. In males, test the cremasteric reflex by stroking the thigh and observing ascent of the testicles. Test anal wink by observing contraction of the anus with light pinprick stimulation. Test the bulbocavernosus reflex by stretching the penis and detecting contraction of the anal sphincter by digital examination. The bulbocavernosus and anal wink reflexes are usually tested in suspected spinal cord injury.

In Conclusion
The neurological examination is directed toward the patient's chief complaint, with emphasis on important areas in the history. In patients with rapidly evolving syndromes, the most important part of the neurological examination is reevaluation and reassessment.

II. NEW ONSET SEIZURES

Take the following steps for a previously undiagnosed patient who presents with a seizure:

• Quickly assess ABCs. Protect C-Spine if trauma is suspected.
• To establish the diagnosis of seizures, statements of other witnesses are essential. Pertinent history includes head trauma, alcohol or drug use, and prior seizures.
• Perform physical examination, including a general exam. Rule out systemic infection and organ failure. Assess postictal confusion and seek focal neurological deficit with specific neurological evaluation. Monitor blood pressure, cardiac, and respiratory functions.
• Assess for continued seizure activity if the patient fails to regain consciousness.
• Provide airway management, administer oxygen via nasal cannula or face mask, or by nasal or oropharyngeal airway.
• Establish a secure IV with an 18 or 20 gauge catheter. Administer normal saline at KVO. Note that Dilantin precipitates in glucose solutions.
• Obtain the following laboratory tests:
   ▪ STAT CBC, electrolytes including calcium and magnesium, EKG, UA, blood sugar.
   ▪ CXR (R/O aspiration), cross table lateral C-spine if history of trauma.
   ▪ Medication screen: Aminophylline, Digoxin, Lithium, anticonvulsants.
   ▪ Drug screen: ETOH, amphetamines, cocaine, barbiturates, PCP.
   ▪ Blood and urine cultures. If septic, obtain Counter Immune Electrophoresis (CIE) antigens.
• Obtain cranial CT Scan without contrast to rule out subarachnoid hemorrhage, and with contrast to rule out tumor and abscess.
• Obtain lumbar puncture if meningeal signs are present without focal neurological deficits; start IV antibiotics early if meningitis is suspected.

- Institute anticonvulsant therapy. Oral loading is usually effective if the patient is not in Status Epilepticus. A single idiopathic seizure may not require therapy.
- Frequently reassess the patient's clinical status.

Approach to status epilepticus
- Quick assessment of general and neurological state. Verify that unconsciousness persists.
- Maintain airway. Use the chin lift/ jaw thrust maneuver.
- Prevent aspiration by placing in semiprone (Fowler) position. Clear secretions, insert airway or intubate, administer oxygen, insert NG tube.
- Start 2 IV lines, 18-20 gauge, administer normal saline at KVO rate.
- Monitor vital signs frequently. Institute cardiac and respiratory monitoring.
- Obtain laboratory tests, CT scan, and lumbar puncture (see above).

Treatment of status epilepticus
- Thiamine 100 mg IM.
- D50W - 1 AMP (50 ml) IV push.
- Narcan 1 AMP IV or via ET tube.
- Valium 5-10 mg IV or via ET tube - Only temporizing measure to assist IV or ET tube insertion. Respiratory depressant, particularly with barbiturates.
- Dilantin (Phenytoin) Note that dilantin precipitates in glucose solutions.
    - Loading dose:
      15-20 MG/KG at 50 mg/min slow IV while on ECG monitor. Observe closely for arrhythmia or hypotension.
    - Maintenance dose:
      100 mg slow IV push q8h to maintain therapeutic levels.
- If status epilepticus persists over 30 minutes administer Phenobarbital 15-20 mg/kg up to 100 mg/min. Follow the patient closely for respiratory depression and hypotension. Consider intubation.
- After 45 minutes, consider Paraldehyde 4%, 5 ml in 5 ml mineral oil IM in buttocks (10 ml per side) or rectally, repeated every 30 minutes as necessary. Note: Paraldehyde is a very messy drug, has a smell, may melt plastic, and is often unavailable.
- Lidocaine 1 mg/kg IV or via ET Tube as loading dose, then 2-4 µg/minute IV. Closely monitor cardiac status.
- If still in status epilepticus after 45-60 minutes, try general anesthesia.
- Obtain electroencephalogram (EEG) as indicated.

III. VERTIGO AND DISEQUILIBRIUM

Introduction
Patients with dizziness, vertigo, and disequilibrium may present with a variety of complaints, including dizziness, lightheadedness, unsteadiness, imbalance, and sensations of spinning, floating, and swaying, to name just a few. History is the most important aspect of the evaluation. Based on the history, dizziness should be classified into one of four types:

- True vertigo - definite rotational sense
- Presyncope/syncope - sensation of impending faint or loss of consciousness
- Disequilibrium - sensation of unsteadiness or loss of balance
- Ill-defined - lightheadedness not otherwise classified

By classifying the patient into one of these four categories, a more pertinent differential diagnosis is established. This is further narrowed by examination and diagnostic testing, which guides therapy and treatment. Despite a through evaluation, an identifiable etiology may not be established. Sensation of dizziness, vertigo, and disequilibrium may be due to:

- Peripheral vestibular dysfunction
- Central vestibular dysfunction
- Systemic dysfunction
- Nonorganic (psychiatric) dysfunction

Thoroughly evaluate the characteristics and pattern of the vertiginous sensation . Establish the rapidity of onset and duration. Identify factors which make the vertigo worse, such as positional changes, or opening or closing the eyes. Associated auditory symptoms, such as tinnitus, ear fullness, pain, or hearing loss usually indicate peripheral vestibular dysfunction. Signs associated with central neurological dysfunction include diplopia, ataxia, dysphasia, dysphonia, and sensory or motor complaints. Important historical factors include prior head injury, recent viral infection, toxic exposure, and medication use.

Physiological Substrates of Vertigo
Spatial orientation is maintained by utilizing sensory information from the visual, vestibular, and somatosensory systems. This information is processed in the brain stem, and finally integrated in the cortical perceptual system. Disruption or alteration of processing of signals from the visual, vestibular, or somatosensory system may cause disorientation or vertigo. For example, a patient who has had cataract extraction may have visual distortion resulting in profound disorientation. A patient with a peripheral neuropathy may have diminished proprioceptive input from joints and muscles, resulting in substantial disequilibrium, particularly in low light situations where the reduction in visual input further degrades orientation.

Vertigo is a hallucination of movement or an erroneous perception of motion, either of self or of external objects. It is usually unpleasant, due to distortion of the static gravitational orientation perceived by the cortical spatial perceptual system. This erroneous perception of motion may be linear or angular (rotatory). This section will focus on the relationship of the vestibular system to vertigo and disequilibrium. The vestibular system has 2 orientation functions:

- Maintenance of postural tone
- Stability of visual-ocular position

The inner ear has two linear motion detectors, the utricle which detects linear motion in the front to back (transverse) plane and the saccule in the side to side (sagittal) plane. These linear motion detectors provide input for postural maintenance by the vestibular system. The vestibulospinal system maintains erect posture and counteracts the effects of gravity on body position. The inner ear also has angular motion detectors, the semicircular canals. Their input enables the oculomotor system to maintain ocular stability, particularly during movement.

Vertigo and disequilibrium can result from mismatch of sensory signals from the static and dynamic spatial orientation systems. There is overlap among the centrally processed visual, vestibular, and somatosensory signals. Central compensatory mechanisms enable deficiencies in one signal to be gradually overcome by other sensory input. As a result of this reprocessing of signals by the central nervous system, symptoms of peripheral labyrinth dysfunction will eventually resolve. Symptoms of central nervous dysfunction, although usually milder, tend to persist. The intensity of vertigo or disequilibrium is related to the degree of mismatch between deficient and intact sensory inputs. Because of the interaction between various inputs during central processing, other symptoms besides vertigo may be experienced. Vertigo itself is perceived at a higher cortical level. Vertigo may be due to either excessive physiological stimulation or to pathological dysfunction.

Gait imbalance, or ataxia, results from abnormal input from the vestibulospinal system. Nausea and vomiting may result from activation of the chemoreceptor trigger zone, or medullary vomiting center. Nystagmus, rhythmic jerking eye movements, may be observed during dysfunction of the vestibulo-ocular processing center in the brain stem or of the peripheral vestibular system.

Physiological Vertigo Syndromes
In physiological vertigo, disequilibrium is due to excessive visual, vestibular, or somatosensory input which is not compensated by other input. In pathological vertigo, there is either an abnormal sensory signal from a sensory receptor, or abnormal signal processing by the central nervous system. Examples of physiological vertigo include motion sickness, space sickness, height vertigo, visual vertigo, somatosensory vertigo, "head extension" vertigo, and "bending over" vertigo. Physiological vertigo has great significance in operational medicine, particularly motion sickness.

Motion Sickness
Motion sickness is due to sensory conflict. We make several assumptions about our visual world. With head movement in one direction, we expect the visual scene to move in the opposite direction. Since we evolved in a one G horizontal plane, we are accustomed to gravitational movements in the horizontal plane only, not in the vertical plane. The semicircular canals sense rotations and the otolith organs detect to-and-fro and side-to-side motion. Motion sickness appears to be worst with movement frequencies from 0.2 to 0.6 Hz. Although infants under age two are quite resistant to motion sickness, they may develop a problem in adolescence or young adulthood. Motion sickness is worsened by removing or altering the surrounding visual

environment. Motion sickness may occur in aircrew who lack an outside visual reference horizon, especially during rapid changes in aircraft attitude or unanticipated flight maneuvers. Motion sickness can be overcome by central adaptation and habituation. Adaptation is aided by anxiety reduction (relaxation techniques, reducing life stress), good hydration, adequate sleep, regular exercise, regular meals, and avoidance of tobacco, caffeine, and alcohol. Aviation personnel who wear contacts should not change them or alternate between contacts and glasses. Doing so may affect the vestibulo-ocular reflex, causing sensory conflict. Aviators who only wear glasses at night may develop motion sickness and disorientation for the same reason. In neophyte aviators, pharmacologic intervention may accelerate adaptation. One of the most effective medications is scopodex (25 mg of scopolamine hydrobromide with 5 mg of dextroamphetamine). Another effective medication is promethazine (25 mg) with ephedrine (25 mg). Pharmacological intervention is a temporary measure. A beneficial effect should be seen within three to five doses. Use medication in conjunction with continued flight training. An airsickness desensitization program is available at NAMI which uses the cross coupled coriolis effect, a potent vestibular stimulus. Balance practice may enhance adaptation to visual vestibular conflict. During balance practice, the patient stands with one foot in front of the other, looking at the ceiling, with arms crosses and eyes closed. This balance practice can be enhanced by standing on one foot, which is extremely difficult. Balance practice enables the patient to become habituated to sensory stimuli without visual input. The extended head position places the otolith organs outside of their normal range of sensitivity and may allow the patient to adapt to sensory conflict. In-flight techniques for managing airsickness include avoidance of hyperventilation, fixing vision on a reference horizon, and inhaling 100 percent oxygen.

Height Vertigo
Height vertigo, a type of physiological vertigo, is visually induced. It occurs when stationary objects in the visual field are distant. Height vertigo usually occurs above a height of three meters and reaches a maximum at 20 meters. Ordinarily, the body constantly corrects for normal amount of body sway using visual cues from stationary objects. The farther away the stationary object, the greater the sway needed to produce a cue. Over time, the height vertigo may progressively worsen, and become a fear of heights with associated psychological symptoms. Height vertigo is worsened by standing, staring at objects moving overhead, such as clouds, and by looking through binoculars, which reduces the peripheral visual field. Height vertigo is reduced by sitting or lying down, or by looking at a nearby stationary object on the same horizontal plane.

Visual Vertigo
Another type of physiological vertigo is visual vertigo, also called optokinetic motion sickness or pseudo-coriolis vertigo. Visual vertigo is induced by changing posture in response to the perceived motion of viewed objects. For example, movie viewers will characteristically turns their bodies in the direction of perceived movement on the screen to attain postural stability. This visual vertigo is quite potent, and can be every bit as disorienting as vestibular vertigo. Conflict between visual and vestibular signals may be the cause of flight simulator sickness.

Somatosensory Vertigo

Somatosensory, or orthokinetic, vertigo is an illusion of movement caused by muscle or tendon proprioceptive input from part of the body. Commonly referred to as "seat of the pants" vertigo, somatosensory vertigo may occur while turning in an aircraft. The gravity vector is increased or redirected off the normal gravitational plane, resulting in an illusion called "the leans." False input from the otoliths may also contribute to this illusion.

Physiological Positional Vertigo

Two positional types of physiological vertigo are "head extension" vertigo and "bending over" vertigo. Physiological positional vertigo may occur when the otolith organs are accelerated beyond their functional range with the neck extended or flexed. This effect is aggravated by removing or alternating visual input such as closing the eyes or looking at moving clouds.

Psychogenic Vertigo

Psychogenic vertigo may result from hyperventilation, and may occur in patients with known psychiatric disease. A patient with psychogenic vertigo may have severe subjective vertigo without associated nystagmus or other physical findings. Severe, incapacitating vertigo may occur during in anxiety attacks or in severe height vertigo (acrophobia). Psychogenic vertigo is treated based on the underlying psychiatric diagnosis. Psychotherapy and desensitization procedures are often useful. A diagnosis of psychogenic vertigo presumes that an organic cause for the vertigo symptoms has been ruled out.

Pathological Vertigo Syndromes

Pathological vertigo results from abnormal sensory input or abnormal central processing. Pathologic vertigo may be either visual, somatosensory, or vestibular. Visual pathological vertigo may occur following cataract extraction. The high plus diopter glasses used to correct for loss of the lens cause a significant alteration in the oculo-vestibular reflex resulting in vertigo. Visual pathological vertigo may also occur in patients who have a substantial difference in visual acuity between the two eyes. Somatosensory pathological vertigo can occur in patients with peripheral neuropathies. The loss of sensory input from the muscle spindles and tendon organs reduces proprioceptive information. Sensory deficits are additive, so a patient with both visual dysfunction and peripheral neuropathy may have more disequilibrium with than with either alone. Vestibular pathological vertigo can be caused by either peripheral labyrinth dysfunction, systemic derangement (such as metabolic, endocrine, or circulatory abnormalities), or central vestibular dysfunction.

CLASSIFICATION OF TRUE VERTIGO

True vertigo is classified into four clinical syndromes depending on whether the vertigo is:

- Paroxysmal or sustained
- Rotational or linear
- Positional or non-positional

The first syndrome, paroxysmal rotational vertigo, occurs in definite attacks. The second syndrome, sustained rotational vertigo, lasts a considerable period and does not occur in discrete

attacks. Neither of these syndromes is positionally induced. The third syndrome is positional vertigo, and is induced or aggravated by positional changes. The fourth is linear vertigo, either a side-to-side or to-and-fro disequilibrium.

Paroxysmal Rotational Vertigo
In children and young adults, rotational vertigo attacks are most likely either benign paroxysmal vertigo of childhood or basilar artery migraine. In adults, consider late life migraine equivalents (vertebral-basilar migraine). In older people with vascular disease, consider basilar artery insufficiency. Other conditions which may occur in acute discrete attacks are Meniere's disease, familial periodic vertigo, and rarely, vestibular epilepsy.

Sustained Rotational Vertigo
Sustained rotational episodes may be seen in Meniere's disease, acute vestibular neuronitis, vestibular nerve lesions (acoustic neuroma), and brain stem lesions.

Positional Vertigo
Positional vertigo may occur in brief attacks or may persist following provocative position changes. Most commonly, positional vertigo is benign paroxysmal positional vertigo (BPPV). However, it may also occur in perilymph fistula, various toxic conditions such as alcohol-induced vertigo, in basilar artery insufficiency, and with central nervous system lesions of the vestibular nucleus or midline cerebellar region.

Linear Vertigo
Linear vertigo results in disequilibrium and postural imbalance, and may be seen with both peripheral or central nervous system pathology. Lateral (side-to-side) imbalance and disequilibrium suggests disorders of the otolith organs, the vestibular nucleus or the lateral medullary syndrome due to vertebral artery occlusion affecting the midline cerebellum. Fore-and-aft postural imbalance occurs in upper brain stem dysfunction, and may be due to a variety of pathological conditions including degenerative, neoplastic, toxic, and vascular diseases.

Specific Vertigo Syndromes
Benign Paroxysmal Positional Vertigo One of the most common peripheral vestibular syndromes is benign paroxysmal positional vertigo (BPPV), which may occur at any age. The characteristic history is brief episodes of positionally induced vertigo, particularly with rapid position changes, such as getting out of bed. The rotational sensation usually lasts less than one minute. However, a nonspecific dizziness or disequilibrium, often described as a swimming sensation, may last hours to days. Although BPPV may remit spontaneously, fully one third of patients have recurrent symptoms over one year. In a review of 240 cases of BPPV, 49% had no identifiable cause established. Eighteen percent occurred following head injury, usually within 3 days. Fifteen percent had a viral illness within the previous two weeks. The remaining 18% had a variety of miscellaneous diagnoses (6% had signs of central nervous system involvement). BPPV symptoms

are often reproduced by performing the Hallpike (or Barany's) maneuver wherein the patient is moved from sitting to supine with the neck extended 45 degrees and the head rotated 45 degrees toward the ground. This maneuver may induce rotational nystagmus when the patient looks toward his "down" ear. An upbeating vertical component may develop when he looks towards his "up" ear. The nystagmus appears after a variable period up to 40 seconds. It usually reaches a peak within ten seconds, then slowly decays over 40 seconds. On repeated Hallpike maneuvers, adaptation develops and the nystagmus gradually lessens. This pattern of nystagmus often reverses direction when the patient reassumes the upright position.

The most likely etiology of BPPV is cupulolithiasis. The utricle is located above the posterior semicircular canal. Calcium deposits, or otoconia, from the utricle degenerate and drop into the sensory portion of the posterior semicircular canal, or cupula. There they land on the hair cells. The hair cells are surrounded by the gelatinous ampulla which has the same specific gravity as endolymph, and normally only detect angular acceleration. When otoconia rest on the hair cells, the semicircular canal is converted from an angular to a linear motion detector, and gravity is perceived as a rotational motion, especially during movements such as the Hallpike maneuver. BPPV is managed with physical therapy. Repeated positional maneuvers promote loosening and dispersion of the fallen otoconia from the hair cells, and permits semicircular canal recalibration by central processing mechanisms. The patient starts in a sitting position and tilts laterally to the lateral decubitus position for at least 30 seconds, or until the vertigo subsides. The patient then sits up for at least 30 seconds, then tilts to the lateral position on the opposite side. This sequence of positional changes is done five times, and is repeated three times a day for at least two to four weeks. This results symptom resolution in a majority of cases. An occasional refractory patient may require surgical resection of the posterior ampullary nerve. Drug therapy for BPPV is generally ineffective, and may delay central recalibration.

Vestibular Neuronitis   Vestibular neuronitis, also called vestibular neuritis or acute unilateral labyrinth dysfunction, presents with acute onset of severe vertigo, with associated postural imbalance, nausea, and nystagmus. This syndrome is more severe than BPPV, has a much more prolonged course, and is not positionally induced. Vestibular neuronitis may occur in epidemics due to a viral etiology, and may be a variant of Bell's palsy affecting the vestibular nerve. This syndrome reduces signal from either the semicircular canals or otolith organs, causing rotational or linear vertigo. Due to the reduced signal, the nystagmus fast phase is directed away from the affected side. There are two effects of body motion. The environment appears to move away from the side of lesion. The postural compensation for this sensation causes past pointing and falling towards the side of the lesion.

Viruses known to affect the auditory, vestibular, and facial nerves include mumps, measles, infectious mononucleosis, and herpes zoster. Herpes zoster can present with ear pain, facial palsy, deafness, and vertigo. It is diagnosed if vesicles are present in the external ear, the Ramsey Hunt syndrome. Management and therapy for vestibular neuronitis depends on the clinical stage. In the first three days, when there is significant nausea and vertigo, put the patient on strict bed rest, with eyes closed and no exercise or head movement. During this phase, antihistamines,

antiemetics, and antivertiginous medications may be useful. Three to five days after the onset of acute vertigo, the patient will probably have spontaneous resolution of nausea and be able to partially suppress nystagmus by fixation. During this phase, aid adaptation with mild exercises in bed such as moving from supine to sitting, practicing fixation on a slowly moving finger, and fixating on a stationary object while the head is slowly rotated in different directions. As improvement is obtained with these measures, the patient may try sitting unassisted. In five to seven days, after resolution of nausea and with only mild residual vertigo, the patient should be able to totally suppress nystagmus by fixation. There may still be nystagmus with fixation removed, using frenzel lenses. At this stage, the patient can try resting on all fours, then advance to resting on both knees. If this is well tolerated, the patient may stand erect with legs spread apart. As symptoms improve, opening and closing the eyes with the neck extended may be attempted. As balance improves, an aggressive eye tracking exercise may be performed by having the patient follow a finger through rapid transitions of gaze or by fixating on an object while the head is rotated back and forth at ever faster rates. Generally, within two to three weeks, all vertigo ceases and even spontaneous nystagmus with frenzel lenses is reduced. At this stage, the patient may try balance walking in the tandem position with the eyes closed and the head extended. Drug therapy is effective only in the first three to five days to reduce the severe vertigo and nystagmus. The overall goal is to allow brainstem compensation mechanisms to readapt to the altered signals. Continued use of medication after five days may actually delay recovery. Exercises using eye, head, and body movement are designed to provoke the sensory mismatch and allow compensation to proceed more rapidly.

Meniere's Disease Meniere's disease, or endolymphatic hydrops, is a common cause of recurrent vertigo and auditory symptoms. It accounts for approximately 10 percent of vertigo cases. Early in the course of Meniere's disease, there is a fluctuating hearing loss in the low frequencies, a sensation of ear fullness or pressure, and unilateral tinnitus which may persist between episodes. Vertigo, postural imbalance, and nausea usually reach a peak over several minutes, and resolve over several hours. There is often a reduced tolerance for loud noises. Early in the course of the disease, the hearing loss is reversible. As the disease progresses, the hearing loss becomes permanent, initially affecting the low frequencies. Late in the course of the disease, vestibular drop attacks due to loss of reflex postural tone may cause sudden falls to the ground. During the vertiginous attack, which usually lasts 30 to 60 minutes, a characteristic nystagmus is seen, with the fast phase away from the affected ear. During the recovery phase following the attack, the nystagmus beats towards the side of the lesion.

The pathophysiologic cause of Meniere's disease is distension of the endolymphatic sac, or endolymphatic hydrops. As the membranous labyrinth progressively dilates and increases pressure on adjacent auditory structures. As the disease progresses there is disruption of otolith organs and semicircular canals, resulting in vestibular symptoms. Dilatation of the membranous labyrinth may lead to rupture of the endolymphatic membrane. This rupture allows endolymph to leak into the perilymph, causing immediate damage to the auditory and vestibular hair cells and nerve fibers.

Distension of the endolymphatic sac may be due to either insufficient fluid reabsorption by the endolymphatic sac or blockage of the endolymphatic duct. Several etiologies have been identified in Meniere's disease. Approximately 50 percent of the patients have a positive family history, suggesting a genetic predisposition. Trauma, infection, or inflammation may block the endolymphatic sac, preventing reabsorption, and leading to endolymphatic sac distension. Thirty percent of patients with Meniere's disease will progress to bilateral involvement. Up to 80 percent may have remission of symptoms, sometimes lasting over five years. However, in some patients the progression of symptoms is quite disabling.

The diagnosis of Meniere's disease is based on the characteristic clinical history. A number of clinical tests have been developed. In classic Meniere's disease, the low frequency hearing loss is reversed by administration of a dehydrating agent such as oral glycerol, which can improve the hearing loss by at least 15 to 20 decibels within one to two hours after administration. Medical therapy is the mainstay of treatment for Meniere's disease. A common regimen includes a low salt diet (800 to 1000 mg of sodium a day) and a diuretic, such as hydrochlorothiazide at a dose of 50 mg QD.

Perilymph Fistula Perilymph fistula may cause episodic vertigo and sensorineural hearing loss. The vertigo is usually not as severe as BPPV. There is a usually a history of trauma or ear surgery. The trauma may be relatively trivial, and can occur after diving, strenuous exercise, exertion, or air travel. Generally, vertigo is precipitated by some type of exertion, valsalva, or position change, and usually last longer than BPPV. The pathophysiology of perilymph fistula is elasticity of the bony labyrinth around the oval or round windows. Because of this elasticity, increases in venous or middle ear pressures are directly transmitted to the membranous labyrinth of the auditory and vestibular apparatus. Fistula testing is designed to increase this pressure. Common fistula tests include compression on the tragus, application of positive or negative pressures or loud noises to the tympanic membrane, and having the patient swallow or valsalva. Exacerbation of symptoms with these maneuvers suggests perilymph fistula. However, vertigo may be induced by valsalva with Chiari brain stem malformation. Other causes of post traumatic vertigo include BPPV and cervical, or whiplash, vertigo. Management of perilymph fistula includes bedrest, head elevation, and administration of stool softeners to avoid valsalva maneuvers. If the symptoms remain disabling for four months, surgery should be considered.

Positional Alcohol Nystagmus and Vertigo Anyone who has "tied one on" can attest to the severe disorientation and vertigo caused by alcohol excess. When blood alcohol exceeds 40 mg percent, it diffuses into the cupula, or hair cell area, faster than into the surrounding endolymph. This causes an imbalance between their respective specific gravities, which turns the semicircular canals, which normally detect angular motion, into linear motion detectors, and makes them susceptible to gravity. Positional vertigo and nystagmus develop. The nystagmus fast phase component beats towards the lower ear. As the alcohol gradually diffuses into the endolymph, it equilibrates. Positional vertigo resolves 3-5 hours after cessation of alcohol consumption. As the alcohol is metabolized, it diffuses out of the cupula more quickly than the endolymph, again causing an imbalance. This occurs five to ten hours after drinking. The nystagmus fast phase now

beats toward the uppermost ear, usually as the blood alcohol level drops below 20 mg percent. This imbalance again causes significant disequilibrium, and the resulting motion sickness is a major component of a hangover. A morning-after drink may temporarily reequilibrate the specific gravity differential between the endolymph and cupola, causing a transient reduction in symptoms. The imbalance may persist 10-12 hours after the last drink; tghus consumption should cease at least 12 hours prior to flight activities.

Toxic Vestibulopathies Toxic substances known to cause vertigo and auditory symptoms include heavy metals and numerous medications. Aminoglycoside antibiotics such as streptomycin and gentamicin are vestibular toxins, neomycin and kanamycin are ototoxic. Other vestibular and ototoxic medications include aspirin (tinnitus is common in therapeutic doses), chloroquine, lasix, quinidine, and quinine, including tonic water. Toxic vestibulopathies may be persistent.

Less Common Causes of Peripheral Vestibular Dysfunction Peripheral vestibular dysfunction may result from diseases of the bony labyrinth, including Paget's disease, otosclerosis, chronic mastoiditis, and congenital or acquired syphilis. Labyrinthine infarction, associated with vascular disease, may cause episodic vertigo. Autoimmune diseases, such as Cogan's Syndrome (episodic vertigo, tinnitus, bilateral deafness, interstitial keratitis, photophobia, ciliary injection, decreased vision) may affect the auditory-vestibular system.

Central Vestibular Vertigo Central causes of vertigo are less common than peripheral or systemic causes. Although lesions of the vestibular nuclei and the vestibular portion of the cerebellum can cause vertigo, other signs of central nervous system dysfunction will be present. The presence of other neurological signs helps distinguish central from peripheral vertigo. Symptoms result from involvement of brain stem structures responsible for facial, extremity, and trunk sensation, eye movement, speech, and facial and extremity motor control. Causes of vestibular vertigo run the spectrum of neurological disease, and include epilepsy, migraine, congenital malformations, and vascular, demyelinating, neoplastic, degenerative, and infectious diseases. Central vertigo tends to be less severe than peripheral vertigo, with fewer autonomic symptoms such as nausea and vomiting. It tends to persist, and is usually less sudden or severe, except for migraine or vascular disease. Evaluation of nystagmus, discussed in the section on vestibular function testing, may help differentiate central from peripheral vertigo.

Miscellaneous Causes of Central Vertigo The Arnold Chiari malformation may cause vertigo when intracranial pressure is increased by maneuvers such as the Valsalva, or when the brain stem is compressed by positions such as head hanging or neck extension. Multiple sclerosis is the great imitator of neurological disease. It accounts for less than five percent of vertigo, but becomes a more likely diagnosis if there is a history of other neurological findings, such as optic neuritis or spinal cord involvement. Various degenerative brain stem conditions may cause in vertigo, and often have a positive family history. Meningitis or encephalitis affecting the brain stem may result in vertigo, usually in association with other cranial nerve and brain stem signs. Extrinsic and intrinsic tumors which may cause vertigo include acoustic neuroma, meningioma, cholesteatoma, chordoma, glomus jugulare tumor, epidermoid tumor, and intracranial metastasis. Intrinsic

tumors of the brain stem and cerebellum, such as glioma and hemangioblastoma, may also result in vertigo. Neoplastic processes usually have a slowly progressive course and rarely cause acute vertigo, unless they hemorrhage or suddenly increase in size.

Vertigo and nystagmus due to the dysfunction of brain stem compensation may persist for a considerable time. Central nystagmus looks more severe than would be expected from the patient's corresponding symptoms of vertigo or nausea. Postural changes tend to stimulate peripheral vertigo more than central vertigo. Peripheral vertigo tends to be reduced with fixation (with the eyes open). Central vertigo tends to be worse with the eyes open, because of the conflict of visual and vestibular information. With eyes closed, the visual information is reduced, which reduces the visual vestibular conflict and vertigo. Peripheral vertigo tends to lessen with repeated head movements, because of brainstem adaptation. In central vertigo, the vertigo may not fatigue or habituate with repeated movements, and nystagmus may vary in direction and amplitude on a daily basis.

Vestibular Function Testing
Clinical tests help establish an etiology in patients with vestibular symptoms. Evaluation of the vestibular system includes a general neurological examination to establish any other nervous system involvement. Routine tests of vestibular function include electronystagmography (ENG) and brain stem auditory evoked response (BSAER). Specific vestibular tests include evaluation of the vestibular spinal reflex, the vestibular ocular reflex, the optokinetic (visual ocular) reflex, station and gait, and provocative tests (posture, position, and fistula testing). The search for spontaneous or positional induced nystagmus is an essential part of this examination.

Vestibular function tests are important to establish nystagmus as central or peripheral. Central causes of nystagmus imply a more serious prognosis, and usually require referral to a neurological center. Other associated neurological findings may aid in establishing the diagnosis. Specialized tests for evaluation vestibular dysfunction, such as the vestibular ocular reflex performance test (VORPET) pendular eye tracking test, platform posturography, and the visual vestibular interaction test (VVIT), are available at the Naval Aerospace Medical Institute/Naval Aerospace Medical Research Laboratory.

Vestibular Function Battery
Vestibulospinal reflex tests include tests of posture, extremity drift, station, and gait. One test of the extremity vestibulospinal reflex is past pointing, which is a reactive deviation of the extremities caused by an imbalance in the vestibular system. To test past pointing, the patient extends his arms and touches his index finger to the examiner's index finger. The patient then closes his eyes, raises his extended arms overhead (vertical position), then returns the index fingers to the examiner's. Damage to the vestibular system causes lateral deviation of the arm and finger on returning to the original position. Accurate past pointing also depends on intact extra-labyrinthine function (i.e., no weakness). With acute peripheral vestibular dysfunction, past pointing occurs toward the side of the lesion. However,after compensation, past pointing causes deviation to the side opposite the lesion. Past pointing is different from the cerebellar dysmetria

(finger to nose) test. Another variant of past pointing is the Quix test. The patient stands, eyes closed, with arms straight ahead. Lateral drift is a positive (abnormal) test.

The Romberg is another vestibulospinal reflex test. There are three Romberg positions, the standard Romberg (feet next to each other), the Tandem Romberg (the patient stands with one foot in front of the other), and the Sharpened Romberg (same tandem stance but with the patient's head placed first straight ahead, then looking at the ceiling). The patient is tested in each position with the eyes open, then closed. Observe any deviation or falling, which is usually toward the damaged side. The sharpened Romberg is very difficult, and may be made even more difficult by having the patient stand on one foot (first dominant, then non-dominant) or by placing the hands on opposite shoulders. Record the best of three times that the patient remains erect. A healthy naval aviator should be able to stand in the Sharpened Romberg position with the head extended and eyes closed for 30 seconds, and on one leg with hands on shoulders for 10 seconds.

The vestibulospinal reflex may also be assessed by the Fukuda Step Test. The patient walks with his eyes closed three steps forward, then three steps backward, for at least 20 cycles. In the absence of cerebellar or proprioceptive dysfunction, deviation or rotation toward one side may indicate labyrinthine dysfunction. The Unterberg step test is conducted in a similar fashion, with the patient marching in place over the same spot. Test tandem gait, looking for deviation.

Vestibular Ocular Reflex Tests The vestibular ocular (VOR) reflex may be tested in a variety of ways. One sensitive test is the dynamic illegible E test. Have the patient read a visual acuity chart while you rotate his head in the horizontal, vertical and lateral (tilt) planes at a frequency of approximately two cycles per second. Start the test with the head and eyes straight ahead, then rotate the head from side to side (testing horizontal canals), and up and down (testing vertical canals). Ordinarily, there should be no more than two lines of decrement on the visual acuity chart. The VOR may also be tested by Barany chair rotation. With the eyes closed, the chair is rotated 10 rotations in 20 seconds. Observe for rotatory nystagmus.

To perform bedside caloric testing, irrigate the external auditory canals with water at 44°C and 30°C. The fast phase of nystagmus will develop away from the side irrigated with cold water and toward the warm water. In peripheral labyrinthine dysfunction, the caloric responses are diminished on one side. In central lesions, the nystagmus is more prominent in one direction than in the other, a directional preponderance.

Optokinetic (Visual Ocular) Reflex Tests The optokinetic reflex test visually induces optokinetic nystagmus. This done by moving an optokinetic tape in both directions, in horizontal and vertical planes. A similar test is performed in the Barany chair by having the patient stare off in the distance as the chair rotates. This induces a full field optokinetic response, which should be tested in each direction.

Position Tests Position tests stimulate eye movements or induce symptoms. The head hanging, lateral decubitus, and Hallpike positions are common provocative position tests. To perform head

hanging, place the patient in a supine position with the head and neck extended backwards over the exam table. Test the eyes both with fixation (staring at an object) and without fixation (using high plus cataract glasses or Frenzel lenses). Next, the patient should be tested in the lateral decubitus position with the ear down, to stimulate positional nystagmus and vertigo. The Nyles-Barany or Dix-Hallpike maneuver is used to stimulate nystagmus and vertigo. Move the patient rapidly from the sitting to the supine position with the head and neck moving from straight ahead to 45 degrees of both extension and rotation, with the eyes directed toward the ground. Perform the maneuver both to the right and left. Observe the eyes for at least 60 seconds for nystagmus. Nystagmus elicited in one direction only is characteristic of BPPV.

Nystagmus Evaluation Begin the evaluation with a description of the type. Nystagmus may be pendular, sawtooth, or exponential, increasing or decreasing. Classic vestibular nystagmus has a sawtooth quality. Pendular or exponential types indicate cerebellar or congenital nystagmus. Note the direction of the fast phase, as well as whether the nystagmus is present while looking straight ahead (primary position) or is brought on by looking in a particular direction (gaze evoked). Characteristically, nystagmus increases in amplitude with gaze in the direction of the fast phase (Alexander's Law). Nystagmus may be horizontal, vertical, or rotatory (clockwise or counterclockwise). Record the specific direction observed, left or right, up or down, clockwise or counterclockwise. In general, peripheral nystagmus tends to be mixed, or horizontal in one direction and rotatory in the other. Pure vertical nystagmus is usually of a central origin, although central nystagmus is often mixed. Nystagmus may be either conjugate (nystagmus beats the same way in both eyes) or disconjugate. Note the delay in onset, or latency, of nystagmus following a position change. Central nystagmus usually starts immediately upon the provocation. Peripheral nystagmus is usually, but not always, delayed. Peripheral nystagmus tends to fatigue on continued evaluation (reduction in amplitude or frequency), or on repeated testing (habituation) due to central compensation. Evaluate the effect of fixation on nystagmus. Eliminate fixation with frenzel lenses. Evaluate nystagmus in provocative positions such as head hanging, lateral decubitus, and the Hallpike maneuver.

Fistula testing uses specific provocative maneuvers (valsalva and tragus compression) to elicit nystagmus or other vestibular symptoms. Substantial vertigo and acute nausea are more common in peripheral lesions. With central lesions, nystagmus is usually prominent, but other symptoms are minimal. In general, peripheral nystagmus is inhibited by fixation. Central nystagmus increases amplitude with fixation, although the velocity of the slow phase may be reduced. Positional nystagmus that lasts over 30 seconds in the provocative position is usually central, however 50 percent of persistent positional nystagmus is idiopathic.

IV. SEIZURES AND OTHER SPELLS

Spells are an abrupt (paroxysmal) disruption of normal interaction with the environment. The differential diagnosis includes a variety of neurological, systemic, and psychiatric conditions. The usual presentation is a sudden onset of alteration of mental status, loss of muscle tone and

posture, or excessive motor activity. The neurological differential diagnosis of spells includes seizures, vascular events (TIA or stroke), atypical migraine (basilar artery migraine), syncope including convulsive syncope, paroxysmal sleep disorders (narcolepsy), intermittent movement disorders (paroxysmal choreoathetosis), myoclonus, essential startle response, metabolic encephalopathies (hypoglycemia), and transient global amnesia. The psychiatric differential diagnosis of spells includes anxiety attack, psychogenic fugue, catatonia, psychogenic amnesia, multiple personalities, depersonalization, episodic dyscontrol, and pseudoseizures.

Evaluation of Spells
History obtained from witnesses is the key to evaluation. The quality of the history depends on the time interval between the event and evaluation. After the event, the neurological examination is usually unremarkable. Evaluate the time course of onset (i.e. whether it came on over seconds or minutes), and any precipitating factors, such as sleep deprivation, alcohol consumption, or hyperventilation. The duration of the event and time of recovery is important. The time of day of the event and its relationship to onset of sleep, time of day, or meals, may also be important. Establish muscle tone and position prior to the event. The level of arousal at the beginning, during, and after the event are important clues to the etiology of the spell. Investigate the patient's overall appearance at the time of the event (pallor, cyanosis), as well as any injuries (bitten tongue, bruises) sustained.

Seizures and Epilepsy
A seizure is an uninhibited sudden discharge from a group of neurons resulting in epileptic activity (neuronal storm or excessive paroxysmal neuronal discharge). Epilepsy is derived from the Greek word meaning *"to seize or lay hold of."* A seizure is a single episode of excessive neuronal discharge and epilepsy is a propensity for recurrent seizures. It is estimated that two to five percent of the general population will have one epileptic seizure during their life and that recurrence could be expected in approximately half of these people. It is estimated that 70,000 new cases of epilepsy are diagnosed each year. The prevalence of epilepsy in the U.S. population of is approximately four million.

Seizures are classified according to type or etiology of the seizure:
- Partial (focal) seizures
- Primary generalized seizures
- Partial seizures with secondary generalization

Primary generalized seizures always involve a alteration of consciousness, and include absence (petit mal), myoclonic seizures, clonic seizures, tonic seizures, tonic clonic seizures, and atonic seizures. Partial seizures are seizures that originate in a focal area of the brain and may or may not propagate to other areas. Simple partial seizures do not alter consciousness. Complex partial seizures, which result in altered consciousness, may begin as a simple partial seizure, or start as a complex partial seizure. A complex partial seizure may or may not progress into a generalized tonic clonic seizure.

Depending on the area of the brain involved, the partial seizure may begin with motor, sensory, autonomic, or psychic phenomenon. Since partial seizures may not always progress to tonic clonic movement or alteration in consciousness, this condition represents one of the most elusive diagnoses in neurology and is frequently misdiagnosed. One of the most helpful points in the partial seizure history is the aura. The patient will often describe the aura as a virtually identical sensation every time. The typical progression of simple partial to complex partial to secondary generalized seizure is as follows:

- An aura
- A cry
- A fall
- The fit, which starts as tonic activity then progresses to clonic activity
- Incontinence

The seizure aura is one of the most important items in the history of partial seizure disorders. Aura means "breeze" in Greek, and literally is like the wind blowing over the patient prior to his seizure. It is often described as a premonition or vague sensation of strangeness. Depending on the area of brain involved, a variety of experiences may be encountered. The patient may feel a vague epigastric sensation, such as an empty, sick, nauseated feeling rising up out of the stomach into the mouth. A variety of affective symptoms have been described including fear, pleasure, depression, eroticism, and rarely anger. The patient may have a feeling of familiarity (*deja vu*), or a feeling of unfamiliarity or depersonalization (*jamais vu*).

A variety of hallucinations may also be experienced. Sensations may be quite vivid, and like all partial seizure auras, are usually very stereotypic. Auras may be described as:

- Formed visual hallucinations
- Auditory hallucinations, such as music (not voices)
- Olfactory hallucinations (unpleasant smells such as burning)
- Gustatory sensations (metallic taste)

Sensory aura phenomenon include tingling, numbness, electricity, or heat. Visual illusions may also be encountered, usually distortions in shape or size of objects. The aura may or may not progress to an alteration in consciousness as the epileptic discharge progresses through adjacent areas of the brain.

Another characteristic feature of the partial complex seizure is the semipurposeful automatism. Automatisms are more or less coordinated semipurposeful involuntary motor activity. They occur during altered consciousness, during or after the seizure, and are frequently followed by amnesia for the event. Some examples of automatism include chewing, swallowing, repetitive vocalization, humming, singing, laughing, mimickery, non-directed anger, blinking, gesturing, wandering, fumbling, fidgeting, or non-directed genital activity.

If a seizure generalizes, there will be an initial tonic phase, which starts as a transient flexion of trunk and extremities followed by a 10 to 30 second period of extension of the head and neck,

axial rigidity, clamping of the jaws, and transient respiratory arrest. Shortly thereafter the clonic phase ensues, with 30 to 60 seconds of convulsive activity, which most people would recognize as a seizure. There may be labored breathing and salivating. As the clonic phase progresses, there is a decrease in frequency and an increase in amplitude of the convulsive movements. The flaccid phase may result in urinary or fecal incontinence. This flaccid phase may last two to 30 minutes, and may be asymmetric (Todd's paralysis) in recovery. The ictal (tonic-clonic) phase of a seizure may be as short as several seconds to as long as eight minutes, but usually lasts one to two minutes.

The postictal phase, heralded by the patient's gradual return to consciousness, may last as short as several seconds to as long as 30-60 minutes and averages about five to 15 minutes. It is this postictal phase (postictal confusion) which is the most helpful historical clue in establishing whether or not someone had a seizure. In general, a person who has lost consciousness because of syncope, even if observed to have convulsive syncopal movements, recovers consciousness fairly quickly upon return of normal blood pressure. The patient with a true epileptic event regains their normal level of awareness much more slowly. Confusion arises when a syncopal patient sustains a head injury, and is dazed and confused. It is absolutely crucial to obtain the history from observers actually present at the event to establish the course of recovery and postictal confusion. Absence (petit mal) seizures are the one exception to postictal confusion in generalized seizures. Absence spells occur during adolescence, last less than 10 seconds, may exhibit a variety of automatisms, but have no substantial postictal confusion. Absence seizures may occur several hundred times a day and commonly present as poor school performance. They may progress to generalized tonic-clonic seizures in adulthood.

Etiology of Seizures
Seizures may be due to vascular, infectious, neoplastic, traumatic, degenerative, metabolic, toxic, or idiopathic causes. The idiopathic category accounts for 40 to 50 percent of all seizures in adults. In the early years, birth trauma, metabolic, infectious, and idiopathic causes predominate in the mid-adult age group trauma, tumor, and idiopathic causes are common. In the older age group tumor and vascular diseases are implicated. Drug induced seizures are usually seen with medications parenterally administered in high doses in a patient with a seizure predisposition or exhibiting some altered metabolism which affects drug clearance (liver or kidney disease). Antibiotics (particularly IV penicillin), antihypoglycemics, antiarrhythmics, antidepressants, (amitriptyline and imipramine), anticholinergics, stimulants (amphetamine), aminophyllin, and lithium have been implicated in seizures. Alcohol related seizures that occur in the acute phase of alcohol consumption are due to the toxic affects of alcohol. Alcohol withdrawal seizures occur 24 to 48 hours after ceasing alcohol consumption. Seizures occurring after three to eight days of abstinence are suggestive of delirium tremens.

Posttraumatic epilepsy (PTE) is divided into early epilepsy, which occurs during the first week after trauma, and late epilepsy, which develops after the first week. Posttraumatic epilepsy is significantly related to the degree of brain injury. In penetrating (missile injuries) the incidence of posttraumatic epilepsy is well over 35 percent, whereas in nonpenetrating (non missile injury) the

incidence is usually less than five percent. Posttraumatic epilepsy usually occurs within the first several years after the traumatic event. Approximately 80 percent of these patients who develop posttraumatic epilepsy will do so within two years of the trauma.

Factors influencing the development of late posttraumatic epilepsy include an early posttraumatic seizure, depressed skull fracture, intracranial hematoma, dural penetration, focal neurological deficit, and posttraumatic amnesia over 24 hours with the presence of a skull fracture or hematoma. Post traumatic anterograde amnesia (PTA) has been implicated as a risk factor for posttraumatic epilepsy. In the absence of a skull fracture or hematoma, amnesia longer than 24 hours is associated with an incidence of epilepsy of only 1.5 percent while amnesia of less than 24 hours has an incidence of epilepsy of less than one percent, implying amnesia without other risk factors may not be as significant a factor in the development of posttraumatic epilepsy as was previously thought.

Pseudoseizures

Pseudoseizures, also called psychogenic seizures or nonepileptic seizures, are a type of behavior which resembles an epileptic event but are voluntary and not due to organic pathology. They may resemble organic seizures. As there are no absolute criteria to make the diagnosis, pseudoseizures are often a diagnosis of exclusion, requiring extensive testing at a specialty center. To make matters worse 10 to 30 percent of patients with pseudoseizures also have organic seizures. It is estimated that 5 to 15 percent of patients with refractory seizures not controlled with medication actually have pseudoseizures.

There are several factors that are helpful in distinguishing a organic seizure from a nonepileptic seizure. Pseudoseizures are generally not stereotypic and usually have bizarre behavior and extreme variation. There may be a family history or past medical history of psychiatric disease. The ictal phase of an epileptic seizure is usually less than 100 seconds while the ictal phase in pseudoseizures is usually over 200 seconds. Eye flutter or eyelid twitching occur during the ictal phase of an epileptic seizure, and are not usually seen in nonepileptic seizures. Epileptic seizures are more common in men, while pseudoseizures are more common in women in the 15 to 35 year old age group. Pelvic thrust movements are usually not seen in epileptic seizures, but are common in pseudoseizures. Generally there is no vocalization except at the very beginning of an epileptic seizure (the cry). Vocalization or interaction with the observer may occur throughout the course of a pseudoseizure. In epileptic seizures, there is usually minimal resistance to eye opening. In the pseudoseizure there is marked resistance to eye opening and the eyes have a tendency to look away from the observer, no matter what direction the observer approaches the patient from. Any injury, such as tongue biting or loss of muscle tone resulting in injury, is uncommon in a pseudoseizure, but may be seen in true epileptic seizures. A prolactin level drawn within 20 minutes is markedly elevated (above 1000 MU/L) in a generalized tonic-clonic seizure, and above 500 MU/L in a partial complex seizure. In a pseudoseizure, however, it is within normal limits. In most cases, patients presenting with recurrent events suspected of being pseudoseizures require video monitoring and referral to a seizure center.

Syncope
Syncope is in the differential diagnosis of spells. Syncope is the sudden transient loss of consciousness and muscle tone due to a sudden impairment of brain metabolism due to a reduction in blood flow, oxygen, or energy substrate to the brain. In most cases the distinction between syncope and seizures is made from the history. Classically, the syncopal patient was in an upright posture and often had a presyncopal sensation (feeling of lightheadedness or loss of vision) prior to the event. Upon losing consciousness, the syncopal patient is flaccid, pale, and sweating and has usually not sustained any injury because the loss of muscle tone was gradual enough to allow the patient to reach the ground without serious injury. The tongue is usually not bitten. Incontinence can be seen with either syncope or seizure, and is usually not diagnostic. Difficulty arises when the syncopal event is associated with tonic clonic muscle activity (anoxic myoclonic jerks). jerks. Myoclonic jerks, seen in syncope, are termed convulsive syncope or anoxic myoclonus, and are likely to occur if loss of consciousness exceeds 15 to 20 seconds. The key to differentiating syncope from a seizure is the recovery of consciousness. Following a fainting spell, blood pressure rapidly returns, and consciousness returns to normal without any period of postictal confusion or disorientation in the syncopal patient, unless the patient sustained a head injury from the fall.

Classification of Syncope
Syncope can be divided into one of four categories. Reflex syncope, called vasovagal syncope in older literature, is the most common type of syncope in the young population. Respiratory syncope, cardiac syncope, and areflexic (paralytic) syncope make up the other categories.

In reflex syncope, a variety of situations may be implicated, such as emotion, anxiety, pain, venipuncture, prostate exam, ocular pressure, micturition, defecation, or postural change. Situational reflex syncope may result from an increased or hypersensitive reflex mechanism.

Reflex syncope is subdivided into vasodepressor or cardioinhibitory syncope. Vasodepressor syncope is due to peripheral vasodilatation of the muscle bed. Cardioinhibitory syncope is due to a increased vagal tone, which slows the heart rate. In vasodepressor syncope, the patient looks pale and feels cold, due to vasoconstriction of the skin and the presence of sweat. There are four physiological phases of vasodepressor syncope. In the presyncopal phase there is a gradual fall in blood pressure and cardiac output. In the compensatory phase there is a gradual increase in heart rate and peripheral vascular resistance in response to the falling blood pressure and cardiac output. Finally, in the syncope phase there is a precipitous drop in peripheral vascular resistance due to vasodilatation of the skeletal muscle bed, resulting in a drop in a blood pressure and heart rate. In the recovery phase, blood pressure, heart rate, and cardiac output increase. and there is a gradual rise of peripheral vascular resistance. Although a variety of precipitating events, such as change in posture, diminished blood volume, anoxia, or fear may trigger vasodepressor syncope, they all progress through these phases. Some situational reflex syncopes such as micturition and carotid sinus syncopes may result from vagal slowing due to a cardioinhibitory response. Vagal (cardioinhibitory) syncope is less common than vasodepressor syncope and may result in syncope even in the recumbent position. Cardioinhibitory syncope has been implicated in cardiac arrest in

athletes and sudden infant death in children.

The next category, respiratory syncope, occurs in a variety of situations, such as coughing, playing wind instruments, or during weight lifting. Respiratory syncope may result from an increase in intrathoracic pressure (over 250 to 300 mm Hg) resulting in an increase in cerebral venous pressure, subsequent elevation in intracranial pressure, and reduced cerebral perfusion pressure. Increased intrathoracic pressure may also cause impaired venous return to the heart, reducing cardiac output. A cardioinhibitory mechanism may result from 1) a transient rise in blood pressure resulting in carotid sinus stimulation, causing vagal slowing of the heart, or 2) hyperactive pulmonary stretch receptors in the lung wall, causing a pulmonary stretch reflex, resulting in cardiac slowing. Cough syncope, called laryngeal vertigo in older literature, occurs in obese males with chronic bronchitis and emphysema and commonly results in a baroreceptor response and vagal slowing. The valsalva maneuver causes elevation in the atrial blood pressure, however it is increased intrathoracic pressure that results in a hyperactive pulmonary stretch reflex and vagal slowing.

The next category is cardiac syncope, which is due to a reduction in blood flow due either to a dysrhythmia or outflow obstruction. Examples of cardiac syncope include the Stokes Adams attack (complete heart block), the sinoatrial node dysfunction (sick sinus syndrome), and the tachycardia bradycardia syndrome, seen in paroxysmal atrial tachycardia and paroxysmal supraventricular tachycardia. Syncope occurring during exercise or exertion may be due to ventricular outflow obstruction from aortic stenosis, or underlying cardiac disease, such as cardiomyopathy.

The final category of syncope is areflexic (paralytic) syncope. Unlike reflex syncope, where a hypersensitive reflex is responsible for the drop in blood pressure, in areflexic syncope there is a loss of the autonomic reflex arch which results in loss of the normal compensatory mechanisms which the body uses in controlling blood pressure. In areflexic syncope the skin remains warm, sweating is present, and the heart rate remains unchanged. In vasovagal syncope the skin initially appears pale, cold, and the heart rate usually drops. The reflex failure in areflexic syncope may be due to preganglionic, ganglionic, or post ganglionic sympathetic fiber damage. Preganglionic damage occurs in Tabes Dorsalis, ganglionic involvement occurs in Shy Drager syndrome and spinal cord injury, and post ganglionic areflexic syncope may occur following sensory neuropathy. With dysautonomic or areflexic syncope, patients are more susceptible to dehydration or drug affects. Drugs which may precipitate syncope include oral diuretics, antihistamines, tricyclic antidepressants, benzodiazepine, ganglionic blockers, barbiturates, and anti-Parkinson medication.

Evaluation of Syncopal Patient
The goals of the syncope evaluation are:

- Establish a precipitating event or situation
- Determine any predisposing factors
- Identify a deficiency in the normal compensatory mechanism

- identify hypersensitive physiological responses

Factors which may predispose or contribute to syncope include inadequate diet, dehydration, fatigue, sleep deprivation, emotional stress, anxiety, underlying infection, excessive caffeine use, alcohol intake, and self medication.

A format for evaluating the syncopal patient includes testing designed to stimulate hypersensitive cardioinhibitory reflexes or to detect deficient compensatory responses. In addition to the physical examination and the syncope test battery, laboratory workup might include a complete blood count, electrolytes, glucose tolerance test, 24-hour holter monitor, echocardiography, graded exercise test, and electroencephalography, depending on the most likely etiology.

V. MANAGEMENT OF COMA AND UNRESPONSIVENESS

Consciousness is a state of awareness and appropriate interaction with the environment. There are two aspects of consciousness which come into play in evaluation of a comatose patient. First is the level of content i.e. mental and cognitive function, and second, level of arousal (i.e. the degree of wakefulness). An alteration or reduction in consciousness is due to either diffuse or bilateral impairment of the cerebral hemispheres (cortex) or dysfunction of the brain stem reticular activating system. Clouding of consciousness implies either an inappropriate content or inappropriate level of arousal. Early in the course of coma, a patient may exhibit alternating excitability and drowsiness, incorrect sensory perceptions, decreased attention span, or misinterpretation of external stimuli. Dementia or senility implies an irreversible loss of cognitive function and memory and is usually seen over a more protracted course although it may be acutely precipitated by other problems such as electrolyte derangement. Delirium is a more agitated state of disorientation where the patient's level of arousal may be increased; content, however, is markedly reduced. This is a common feature of toxic and metabolic encephalopathy, such as drug overdose, major organ failure, severe head injury, systemic infection, or subarachnoid hemorrhage.

The degree of drowsiness is often misrepresented on the patient's record. The terms obtundation, stupor, and coma are often used interchangeably. It is best to note the response the patient makes with his environment (i.e., responds to soft verbal stimuli, loud verbal stimuli, physical stimuli such as shaking, or deep painful stimuli to the extremities). Coma or absence of arousal to any external stimuli is mimicked by several other clinical conditions which may be confused with coma. These conditions include:

- Locked in syndrome
- Psychogenic coma
- Persistent vegetative state
- Akinetic mutism
- Hypersomnolence (exaggerated sleep response)
- Brain death

Locked in syndrome is seen in brain stem infarction, metabolic conditions which cause paralysis of all four extremities without loss of consciousness, or acute motor paralysis due to peripheral nerve or neuromuscular junction blockade. There may be preservation of eye movements and blink reflex. Communication may be established by eye blinks. Psychogenic coma should be considered if the patient has intact brain stem reflexes, including caloric nystagmus, pupillary reactions, and optokinetic nystagmus. In psychogenic coma, there is an active resistance to eyelid opening. The patient will tend to avoid looking at the examiner. Persistent vegetative state resembles coma. This condition occurs from severe injury to higher cortical structures resulting in a total lack of response to the external environment, however the patient may still have sleep and wake cycles and spontaneous eye opening. Akinetic mutism results from damage to specific areas of the frontal or limbic cortex, resulting in a loss of interest in the environment, even though the patient may appear otherwise neurologically normal. Excessive sleepiness (hypersomnolence) conditions may mimic comatose states. Brain death may also mimic coma. Nonpsychiatric (organic) coma may be due either to structural, metabolic, or toxic conditions. Structural lesions may involve the supratentorial or infratentorial regions. A history of drug abuse, headache, fever, or previous medical condition might be significant. The patient may not be
able to provide a history, so much of the evaluation will depend on the examination and diagnostic tests.

Physical examination
The general physical examination should include the vital signs. Evaluation of the skin may reveal needle tracks, cyanosis, dehydration, rash (meningococcal infection), or uremia. Bullous skin lesions may occur from drug effects (barbiturates, carbon monoxide, phenothiazine, imipramine and meprobamate). Breath may reveal alcohol, acetone, hepatic failure, or uremia. Cardiac examination may be helpful in finding a murmur, suggesting endocarditis or arrhythmias, which may result from subarachnoid hemorrhage or a brain stem lesion. Hypothermia may be due to exposure, overwhelming sepsis, drug effect, hypoglycemia, hypothyroidism, or Wernicke's encephalopathy. Altered ventilatory patterns may be indicative of metabolic acidosis or respiratory alkalosis. The neurological examination should include a general assessment of consciousness, including response to voice and painful stimuli. Cranial nerve evaluation is important, particularly the pupillary light reflex. It is important to use a bright light when evaluating pupillary responses. Local eye trauma, cataracts, or eye surgery may alter the pupillary response. Preservation of the pupillary light reflexes suggest metabolic coma. Atropine (given following cardiac arrest), amphetamine intoxication, and postanoxic may cause fixed and dilated pupils. A fixed midposition pupil may be seen with hypothermia or glutethimide. Small, fixed pupils may be seen with opiates, organophosphates, pilocarpine, phenothiazine, and following respiratory arrest from barbiturates. Brain herniation may result in fixed pupils even though the herniation may be a primary metabolic process such as cerebral edema. The position of the eyes in their primary resting position should be recorded and whether they are conjugate or disconjugate. Abnormal deviation (include horizontal or vertical), and spontaneous eye movements (roving eye movements, bobbing, or nystagmus) should be evaluated. Assessment of brainstem reflexes should include the corneal reflex, gag reflex, sternutatory reflex,

oculocephalics, and vestibular reflexes. Motor function testing should assess spontaneous movements, such as myoclonic jerks, posturing, asterixis, or seizure activity, or if response to stimuli is appropriate, purposeful, or nonpurposeful.

Some general rules apply in the comatose patient. Usually focal neurological signs indicate a structural lesion, however focal signs may be seen in Todd's paralysis following a generalized seizure disorder or if there is a preexisting focal deficit such as an old stroke. Nonfocal neurological signs usually indicate toxic or metabolic coma, however nonfocal signs also occur in subarachnoid hemorrhage, bilateral subdural hematoma, or vasculitis. A fluctuating neurological examination usually indicates a toxic or metabolic coma, but may also be seen in fluctuating intracranial pressure elevation or status epilepticus (during the refractory or twilight phase). Toxic or metabolic coma usually has an incomplete and symmetric affect on the nervous system, affecting many levels of the neuraxis simultaneously while retaining integrity at other levels. In metabolic coma there is no regional (focal) anatomic defect as in structural coma. Toxic and metabolic coma generally does not cause impairment of horizontal and vertical vestibular ocular reflexes (Doll's eyes). Respiratory patterns may localize the level of the neurological lesion. Damage to the cerebral hemisphere may result in "Cheyne-Stokes" respiration, a hyperventilation pattern with a crescendo- decrescendo amplitude. Damage to the midbrain and higher brain stem structures may result in central neurogenic hyperventilation, which is a hyperventilatory pattern in excess of 20 respirations per minute without the crescendo amplitude seen in Cheyne-Stokes respiration. Damage to the midbrain or pons may cause apneustic or cluster breathing, resulting in a prolonged pause following inspiration. Finally with damage to the medulla in the lower brain stem region, ataxic breathing, similar to a hiccup pattern, may be seen. Hiccups often imply an impending neurological crisis involving the lower brain stem (medullary chemotactic trigger zone). Respiratory patterns suggest involvement at certain levels but are not always diagnostic.

Laboratory Assessment

A screening laboratory evaluation may aid in establishing the cause of coma. Neurological effects often outlast metabolic electrolyte derangement. Evaluation should include complete blood count, electrolytes, arterial blood gases, toxin and drug screens. The electroencephalogram, CT scan, and lumbar puncture may also aid in diagnosis. Treatment for coma should include:

- Initial establishment of the ABCs
- Thiamine 100mg IV push
- Dextrose 50 ML of D50W
- Narcan 2 amp IV push

Once the patient is stabilized from a circulatory and respiratory standpoint, signs of impending herniation syndromes should be sought. If a herniation syndrome is present the patient should be treated for intracranial pressure elevation. Treatment of coma is dependent upon establishing the etiology. The diagnostic tests described above are useful in establishing the appropriate cause. As with all evolving neurological crises, it is extremely important to continually reassess the patient with serial examinations.

VI. CENTRAL NERVOUS SYSTEM INFECTIONS

Introduction

A variety of organisms may infect the central nervous system, often with life threatening consequences. CNS infection may result from viral, bacterial, fungal, protozoal, or rickettsial organisms. Before central nervous system infection can occur, the organism must penetrate extra neural structures, overcome local defense mechanisms, cross the blood brain barrier, then persist and reproduce despite host defenses. Organisms may gain access via direct penetration of the skin (following trauma or surgical procedures) by spread from adjacent cranial sinus or bone infections, by uptake via the peripheral nerve axonal transport system from wounds (rabies, tetanus, or Simian B monkey virus), or by directly penetrating the olfactory mucosa. Most organisms gain access to the central nervous system via hematogenous (blood-borne) spread.

Acute Bacterial Meningitis

The most common bacterial infection of the central nervous system is acute pyogenic meningitis, which is life threatening. Bacterial meningitis was first described in 1805. The first therapy occurred with the advent of lumbar puncture. Intrathecal antiserum was injected via lumbar puncture in 1913 by Flexner. This reduced the mortality of bacterial meningitis from 90 to 30 percent. With the advent of antibiotics in the 1930s, mortality dropped to 14 percent. Despite the improved antibiotics available today, overall mortality rate for acute pyogenic meningitis remains about the same. The pathogenesis of meningitis depends on a defect in the blood brain barrier, bacterial virulence factors, and host defense factors. The type of micro-organism in meningitis is related to patient age, presence of underlying medical conditions, and predisposing factors in the host. Bacterial meningitis is a dynamic process, involving central nervous system penetration, unimpeded bacterial multiplication in the spinal fluid, a secondary bacteremia, and finally a continuous reseeding of the intracranial spaces. Meningitis may alter the blood brain barrier permeability and result in other sequela such as venous thrombosis and brain edema (vasogenic, cytotoxic, and interstitial). Bacteria have developed factors which enhance their survival and facilitate penetration into the nervous system. Perhaps the most striking example is the protein coat of the bacteria capsule which is present in the four major bacterial pathogens: *S. pneumoniae, H. influenzae, N. meningitidis,* and *E. coli.* This encapsulation antagonizes phagocytosis by the white blood cells.

In the neonatal period the primary bacteria involved in meningitis are the gram negative rods *(Escherichia coli)*, and group B *streptococcus*. In infants over three months of age, *Hemophilus influenza* becomes the leading cause. Maternal placentally transferred antibodies protect the infant from *H. influenza* in the immediate post natal period. After three years of age, *H. flu* drops in incidence, and *Streptococcal pneumoniae* and *Neisseria meningitis* become the most frequent pathogens. A variety of medical and surgical conditions may predispose the patient to bacterial meningitis. An immunocompromised state or debilitation, such as chronic alcoholism, may predispose a patient to *Hemophilus influenza, Streptococcus pneumoniae, and Listeria monocytogenes*. Burn patients are more susceptible to *Pseudomonas*. Patients with splenic dysfunction or sickle cell disease are predisposed to *Streptococcus pneumoniae* and *Hemophilus influenza*. Chronic sinusitis may predispose the patient to anaerobic *Streptococcus, S.*

pneumonia, and gram negative rods, such as *Bacteroides fragilis.* Penetration of the skin and dura following post traumatic spinal fluid leak or neurosurgical procedures predisposes a patient to *S. pneumoniae, Staphylococcus aureus,* and gram negative meningitis. A patient with subacute bacterial endocarditis may develop *Staphylococcus epidermitis* meningitis.

Bacterial meningitis in a patient with an underlying medical condition will have a more profound effect on central nervous system function, often resulting in a decreased level of consciousness. Septicemia, overwhelming fever, and deteriorating vital signs are common manifestations of the big three bacterial meningitis organisms: *S. pneumonia, H. influenza, N. meningitidis.* Rash and petechiae are common in *N. meningitidis* but may also present in *S. aureus, Hemophilus influenza, Streptococcus pneumoniae*, or viral meningitis (Coxsackie Echo 9). Signs of meningeal irritation, such as nuchal rigidity, fever, photophobia, headache, and pain on eye movement, may not be present in a infant or child, or in an immunocompromised or elderly individual.

An early diagnosis is crucial and the diagnostic procedure of choice is lumbar puncture and spinal fluid analysis. It is important not to delay antibiotic therapy while waiting for a lumbar puncture or other diagnostic studies to be performed. Cerebrospinal fluid findings in bacterial meningitis include
- Elevated white blood cell (WBC) count, particularly > 1000 WBCs/ cubic mm and > 50% polymorphonuclear neutrophils (PMNs)
- Elevated spinal fluid protein > 50mg%
- Glucose level < 2/3 of simultaneously obtained serum glucose level

Early identification of the responsible organism will aid in the appropriate selection of antibiotics. Bacterial culture and sensitivity assay is essential for guiding antibiotic therapy. The CSF gram stain may provide an immediate clue to the etiology while the culture and sensitivities are pending. Counterimmune electrophoresis (CIE) provides early identification of the common bacterial pathogens *(H. flu, N. meningitidis, and S. pneumococcus)* within hours. Serum, urine, and spinal fluid CIE levels should be obtained. Failure to grow or isolate an organism may be due to prior antibiotic use (often as self treatment for a presumed cold), meningitis due to a nonbacterial infection (fungal, viral, protozoal, Rickettsial), an unsuspected bacterial infection (Lyme disease, tuberculosis, or syphilis), or, lastly, a parameningeal infection (subdural empyema or brain abscess). Every effort should be made to diagnose these conditions, particularly if the patient deteriorates or fails to improve after the administration of broad spectrum antibiotics. If a bacterial meningeal infection is suspected, it is crucial that antibiotic administration not be delayed while diagnostic tests are performed. In severe life threatening sepsis and meningitis with cerebral edema, the patient may need intubation, intracranial pressure monitoring, and treatment of intracranial hypertension. Hyperthermia should be aggressively treated.

Treatment of Bacterial Meningitis.
Community acquired bacterial meningitis in a previously healthy adult will usually respond to penicillin. With the extensive use of antibiotics, bacteria have become increasingly resistant to commonly administered antibiotics. Penicillin resistance in *S. pneumonia* and *Neisseria* is

increasing as a result of a viral plasmid which carries the enzyme, beta-lactamase, which disrupts the antibiotic structure, rendering it ineffective. In *H. influenza* ampicillin resistance is present in approximately 25 percent of cases. One percent are resistant to chloramphenicol. The most appropriate antibiotic is determined by the bacterial sensitivity to antibiotic minimum inhibitory concentrations of less than 0.1 mg/ml. For meningitis of unknown etiology broad spectrum antibiotic coverage is indicated. IV antibiotic therapy should continue for at least 7 to 10 days following the return of normal temperature and clinical stability. Repeat spinal fluid analysis may be indicated within 2 to 3 days if the patient deteriorates. Followup spinal fluid analysis after completion of an antibiotic course may also be indicated if the patient relapses.

Third generation cephalosporins are becoming increasingly popular because of their broad coverage and the emergence of penicillin resistant organisms. Gram negative meningitis may be found in septic urinary tract infections, penetrating head injury, or following neurosurgical procedures. Third generation cephalosporins in combination with aminoglycosides are effective against gram negative meningitis. Intrathecal antibiotics are occasionally indicated for gram negative meningitis, such as hospital acquired *Pseudomonas* in an elderly, debilitated patient. Patients with neurosurgical shunts should have them tapped for spinal fluid analysis. If CSF infection is present, removal of the shunt may be necessary. Patients allergic to penicillin may require erythromycin, chloramphenicol or a cephalosporin.

Prophylactic treatment with rifampin is indicated for *Neisseria meningitidis* for all close contacts of the index case, such as household members, workers, shipmates, or squadron mates who are in close contact, or close contacts in infant day care centers. Casual contacts do not need to be treated. The secondary attack rate for close contacts is about 1%, and is higher for younger children. All contacts should be treated simultaneously. Throat cultures are not effective in deciding who should receive prophylaxis. In adults, rifampin is given at a dose of 600mg q 12 hours for a total of four doses. Minocycline may be effective but causes substantial vestibular reactions. Chemoprophylaxis for *Hemophilus influenza* exposure depends on the age of close household contacts. If the close contacts are children less than 4 years of age in the household of the index case then all household members should receive rifampin (20mg/kg/d dose for 4 days). Infants in day care centers may be considered close contacts in some situations and therapy should be started as soon as possible within 7 days of discovery of the index case. After 7 days, the use of chemoprophylaxis with rifampin is not effective. If the index case and close contacts are over 4 years of age then chemoprophylaxis is not indicated.

Parameningeal Infections
Sinusitis may erode through the dura and may result in meningitis, osteomyelitis, epidural abscess, subdural empyema, subdural abscess, brain abscess, or venous sinus thrombosis. The most common organisms are *S. pneumonia, Streptococcus, Staphylococcus,* and *H. influenza.* Parameningeal infection is a life threatening condition and may be more serious than acute bacterial meningitis. Depending on which sinuses are involved, there may be a variety of clinical presentations. Mastoid sinusitis and involvement of the lateral aspect of the petrous portion of the temporal bone may result in a brain abscess with focal neurologic deficits, seizures, and signs of

increased intracranial pressure (headache, vomiting, and decreased level of consciousness). Sphenoid sinusitis may present with septic thrombophlebitis and cavernous sinus thrombosis, which may involve the optic nerve (visual loss), the trigeminal nerve (facial numbness), or the oculomotor nerves (double vision). Frontal sinusitis and skull osteomyelitis may cause Pott's Puffy Tumor, resulting in a unilateral or occasionally bilateral swelling of the orbital region due to a subperiosteal abscess. Occasionally, the infection extends into the epidural region. A dural tear from previous head trauma may result in a subdural empyema, resulting in rapidly progressing neurologic deterioration, meningeal signs, focal neurologic deficits, and seizures. Treatment is dependent on the responsible organism. The organism usually comes from the adjacent sinus, and is often penicillin resistant *S. aureus* or a gram negative rod. Subdural empyema requires prolonged (2-4 weeks) IV therapy with a penicillinase resistant penicillin, such as nafcillin (12 gm per day), chloramphenicol, or an aminoglycoside. An abscess in the subdural or intracranial space should be surgically treated, the organism identified, to identify the organism, and institute appropriate antibiotic therapy, and to reduce the mass effect.

Nonbacterial Infections
Nonbacterial organisms may involve the sinuses, causing acute neurologic deterioration. In diabetics and leukemics, molds, such as *Rhizopus* and *Mucormycosis,* may result in a fulminant meningo-encephalitis, progressive neurologic deterioration, cranial nerve palsies, seizures, and infarction. Therapy for fungal brain infection is IV amphotericin B. Mortality is very high. Malignant otitis externa is seen in diabetics who develop *Pseudomonas* cellulitis, which spreads intracranially, resulting in meningitis or meningo-encephalitis. The protozoan *Naegleria* may cause a fulminant and usually fatal meningoencephalitis following swimming in infected fresh water. Treatment with amphotericin B and miconazole is a last ditch effort. Spinal fluid analysis in *Naegleria* meningoencephalitis reveals a polymorphonuclear pleocytosis, occasional eosinophils, and mobile ameba.

Systemic fungal infections are common complications of acquired immune deficiency syndrome (AIDS). Four drugs available for treating systemic fungal infections are Amphotericin B, flucytosine, miconazole, and ketoconazole. Rickettsial infections, such as Rocky Mountain Spotted Fever and scrub typhus are treated with a tetracycline or chloramphenicol. Lyme disease, caused by a bacterial spirochete, is effectively treated with tetracycline or penicillin.

Cerebral malaria is a life threatening complication of infection with Plasmodium falciparum. Cerebral malaria is characterized by profound mental obtundation, psychosis, seizures, and hyperreflexia. The cerebral spinal fluid shows an elevation of pressure and protein but no pleocytosis. Fourteen days following the mosquito bite, the patient develops prodromal chills, spiking fever, which progresses to intense headache and muscle pain. The pathogenesis of cerebral malaria is a mechanical distortion of the blood vessels due to rapid proliferation of the parasites, causing stagnation of blood, and possibly a toxic effect on the vascular endothelium or an immune complex vasculitis. Treat cerebral malaria with intravenous quinine. Glucocorticoids, used to treat cerebral edema, have been shown to prolong the coma and increase complications without affecting mortality and are now contraindicated in cerebral malaria.

Viral CNS Infections

Viral infections of the nervous system produce three classic clinical syndromes: meningitis, encephalitis, poliomyelitis. Viral meningitis produces a milder clinical syndrome than bacterial meningitis. The patient has a mild headache and less prominent meningeal irritation. Spinal fluid analysis reveals a elevated white blood cell count that is predominantly lymphocytic. The spinal fluid protein usually remains within normal limits and the spinal fluid glucose remains within two thirds of serum glucose. Viral encephalitis is divided into two categories, infectious encephalitis, due to the direct effects of the virus, and parainfectious encephalitis, due to associated reactions in immune system affecting the central nervous system. Para-infectious complications include a perivenous inflammatory response and leucoencephalitis. Other post infectious syndromes include cerebellar ataxia, and peripheral nerve disorders, such as Guillain-Barre syndrome. Viral encephalitis is classified as arbovirus (arthropod borne), enteroviruses, childhood viruses, and other.

The most common sporadic (nonepidemic) viral encephalitis is herpes simplex encephalitis. This form of viral (HSV) encephalitis may result from either exogenous infection (entrance via olfactory mucosa) or from dormant (latent) virus residing inside the host nervous system (trigeminal sensory ganglion). The characteristic clinical course for herpes simplex encephalitis is an acute or subacute syndrome of headache, fever, behavioral disturbance, seizures and progressive cortical dysfunction. Herpes simplex encephalitis causes a necrotizing hemorrhagic encephalitis, primarily involving the frontal, temporal, and limbic lobes. Spinal fluid analysis reveals red blood cells due to brain hemorrhage and necrosis. Oligoclonal bands on immunoglobulin protein electrophoresis may be present in the spinal fluid. EEG will reveal periodic spike and slow waves over the temporal lobes. CT scan reveals bitemporal necrosis and hemorrhage of the frontal and temporal region. This mass effect is usually present within the first 5 days of onset. Temporal lobe biopsy is the diagnosis of choice and reveals characteristic intranuclear inclusion bodies. Treatment of HSV encephalitis is with intravenous acyclovir.

Epidemic encephalitis is usually related to vector spread, such as insects. The most common arthropod-borne virus is St. Louis encephalitis. St. Louis encephalitis may present with seizures, tremor, myoclonus, vertigo, or electrolyte imbalance. Arboviruses tend to occur during the summer months. The most fulminant of the arboviruses is Eastern Equine Encephalitis, which affects horses and pheasants prior to spread in man. Arboviruses tend to affect children more than adults. Encephalitis due to mumps and lymphochoriocytic meningitis occur primarily in the winter months. Except for herpes virus, there is no specific antiviral therapy other than symptomatic treatment of fever and anticonvulsant therapy if seizures are present.

VII. HEADACHES

Introduction

Headache is one of the most common complaints that plague mankind and one of the most common symptoms seen by a neurologist. Every effort should be made to categorize the headache into a syndrome, and establish the likelihood of an organic or life-threatening cause.

Pain sensitive structures implicated in headache include blood vessels of the scalp and skin, cerebral blood vessels of the skull base (large intracranial sinuses and intracranial arteries), the dura (including the falx), the sensory cranial nerves (V,IX,X) and the upper cervical nerves. The brain parenchyma itself is insensitive to pain. Mechanisms of pain in headache include traction, inflammation, or noxious stimulation of pain sensitive structures, distension or dilation of pain sensitive blood vessels, pressure on cranial or cervical nerves, or contraction of the cranial or cervical muscle bed. An ad hoc committee was formed in 1962 to standardize and classify headaches. They developed a classification scheme based on 15 possible headache categories. A more practical approach to headache classification divides headaches into one of three categories: (1) vascular, (2) tension (muscle contraction), or (3) traction/inflammatory headache.

Approach to Headaches
In approaching headache it is important to ask three questions:
- Does this headache fall into a clinical syndrome?
- Does this headache represent a sign of a life threatening medical condition?
- What impact does this headache have on operational safety?

Of the three clinical headache syndromes, the traction/inflammatory headache is the most likely type to represent a serious medical condition. Factors suggestive of a traction/inflammatory headache include associated loss of consciousness, sudden onset of severe incapacitating headache, associated focal neurological signs, meningeal signs (stiff neck, photophobia, pain on eye movement), altered level of alertness or cognition, change in personality, or associated medical condition such as hypertension or endocrine disease. A headache associated with effort or position change, a change in headache pattern, a headache which no longer responds to treatment, or a headache in a person over age 50 may represent a serious headache. Immediate hospitalization or referral to the appropriate consultant would be indicated if there was an associated recent head injury, focal neurological deficit, sudden onset of severe headache, altered level of consciousness, papilledema, fever, hypertension, or headache in pregnancy.

The Headache History
History is very important since physical signs are rarely evident. The LEARN-IT mnemonic is useful in obtaining a history.

L Location
E Exacerbating factors
A Alleviating factors
R Radiation
N Nature
I Intensity
T Timing

L is for Location Vascular headaches tend to be unilateral in the distribution of a blood vessel.

The location for muscle tension headache is usually band-like around the front and back of the head or the suboccipital region. Traction-inflammatory headaches tend to be retro-orbital or diffuse. Although 2/3 of migraine headaches are unilateral, the possibility of an intracranial mass must be considered if recurrent headaches are always localized to one side. Although migraine headaches may preferentially affect one side, they will occasionally alternate sides. Intracranial lesions may cause a unilateral headache if there is traction on blood vessels or dura, or may be diffuse if there is obstruction of cerebrospinal fluid pathways. Headaches in an elderly patient, particularly if unilateral, throbbing, or associated with neurological findings, may be due to cerebral vasculitis or temporal arteritis, and necessitate an urgent neurological referral.

E is for Exacerbating Factors Exacerbating factors precipitate, aggravate, or worsen a headache. Such factors might include stress, certain foods, bright lights, etc.

A is for Alleviating Factors Alleviating factors reduce or terminate the headache, and might include rest, medication, a dark room, etc.

R is for Radiation This refers to spread after the onset of headache (i.e., where the headache progresses to).

N is for Nature The nature or character of a headache will help classify it as vascular, tension, or traction/inflammatory. Vascular headaches tend to be throbbing in nature, tension headaches tend to be characterized by a band-like sensation of pressure and traction/inflammatory headaches tend to be characterized by deep aching pressure, although they may also be stabbing, sharp, or dull.

I is for Intensity Headaches should be rated on a scale of one to ten with ten being the worse and one the least. The patient should give a value for the average headache and the maximum headache, and relate the intensity of the headache to time from onset.

T is for Timing A time intensity curve may help categorize the type of headache. Vascular headaches tend to build over several minutes to an hour. In the classic migraine vascular headache, the intensity builds to a maximum in 20-30 minutes. Tension headaches tend to increase slowly over hours to days. Traction/inflammatory headaches, depending on the type of pathology involved, may develop over a very short or very long period. The subarachnoid hemorrhage headache usually has a very acute onset over seconds, being described as a lightning bolt headache. Meningeal irritation headache may develop over days. A tumor headache may develop over weeks or months. The time of day or day of week of onset may be important. Headaches that occur early in the morning or awaken a person from sleep may be suggestive of a serious, possibly life threatening headache; however vascular cluster headaches commonly occur at night and awaken the patient from sleep. Tumor headaches tend to be worse with position change and are worse on awakening. Tension headaches tend to be worse on weekdays and are reduced on weekends, and usually intensify as the day progresses. Headaches due to caffeine withdrawal occur on weekends.

Clinical Evaluation

Most patients with headaches will have a normal physical and neurological examination. The examiner should pay attention to the head and neck, including inspection and palpation of the scalp, sinuses, and cervical spine for tenderness. The routine neurological examination should include an evaluation for any signs of neurological deficit, particularly focal neurological findings. A number of ancillary tests may be obtained in the evaluation of a headache patient, including blood count, chemistries, urinalysis, serology, and vasculitis screen. Radiographs of the sinuses, orbits, and temporal bone may be indicated. Clinical studies, such as electroencephalography, visual fields, and evoked potentials may also be helpful in identifying focal neurological abnormalities. Structural workups such as computerized axial tomography (CAT) or magnetic resonance imaging (MRI) may be indicated if neurological findings are present, there is a history of trauma, severe headache, or a traction/inflammatory headache. Lumbar puncture and spinal fluid analysis is indicated if meningeal inflammation is suspected.

Muscle Contraction Headache

Tension or muscle contraction headaches account for over 40 to 50 percent of the headaches seen in a general neurology setting. The tension headache is usually described as an ache, tightness, pressure, crushing, or band-like constriction, varying in intensity, frequency, and duration. These headaches often last days, and are commonly located in the frontal and suboccipital region. These headaches may be as severe or incapacitating as the vascular or migraine headache. The classic muscle contraction headache begins in late morning and progresses with intensity over the afternoon and evening and tends to be worse on weekdays. An intense muscle contraction headache often runs from the suboccipital region down to the shoulders. The most reliable features of muscle contraction headache are the sensation of tightness, pain in back of the neck, pain that intensifies as the day progresses, and pain and muscle contraction associated with anxiety or tension. Depression may also be a feature of this headache syndrome. Other medical conditions, such as cervical spondylosis or nocturnal chewing (bruxism), may contribute to muscle tension headaches. A number of chemical substrates, such as bradykinin or prostaglandins, may aggravate muscle tension, and have been implicated in the headache associated with systemic illness. Tension headache may result from occupational conditions, such as prolonged sitting over a desk, looking at video display terminals, or in air combat maneuvering if the head was extended and rotated off axis while sustaining high Gs.

Treatment for tension and muscle contraction headaches should include elimination of possible stress factors, or other aggravating conditions. Medication for treatment of muscle contraction headaches includes aspirin, acetaminophen, nonsteroidal anti-inflammatory drugs, and, when indicated, muscle relaxants. Therapy with tricyclic antidepressant compounds, such as amitriptyline, may also be helpful, particularly in mixed headaches. The compound Fiorinal, which contains aspirin, phenacetin, butalbital, and caffeine, may also be effective.

Posttraumatic Headache

Ten to forty percent of patients with minor head injury may have an associated headache, particularly with scalp, skull, or sinus involvement. Pain may be steady, cap like, or superficial

over the impact site, and may be aching or throbbing. Prominent neck injury may also be a factor. Headache after head injuries may be due to injury to the scalp vasculature or muscles, or stimulation of small nociceptive pain fibers. Posttraumatic headaches are often associated with imbalance, personality changes, or concentration difficulties. These headaches usually resolve within several months. Persistent posttraumatic headaches may be the basis medicolegal situations or accident compensation. Treatment generally includes analgesics and nonsteroidal anti-inflammatory medication. Resolution of any litigation is essential, as this inevitably contributes to the symptoms.

Vascular Headaches
Vascular headaches are associated with a cerebral blood vessel, either intracranial or extracranial. These headaches are often unilateral and are characteristically pounding or throbbing. Vascular headaches can be divided into nonmigraine vascular headaches and migraine vascular headaches.

Nonmigraine Vascular Headaches
Nonmigraine vascular headaches may be associated with a variety of medical, environmental, and physical conditions which may precipitate a throbbing unilateral headache. Medical conditions associated with headaches include cerebral vascular disease, hypertension, seizure, or endocrine dysfunction (hypopituitarism, hypothyroidism, pheochromocytoma, Addison's disease). Environmental or physical factors, such as hypoxia, anemia, or high altitude, may precipitate a vascular headache.

Altitude Headache/Mountain Sickness
Mountain sickness is often accompanied by severe pounding headache with associated nausea and dimness of vision. The altitude headache is usually a throbbing vascular headache, often generalized and more evident over the frontal areas. It is unusual at altitudes below 8,000 feet, and is an almost universal feature over 12,000 feet in nonacclimatized individuals. It usually occurs in mountain climbers, but may occur in those flying in unpressurized aircraft above 12,000 feet. The headache is not due to hypoxia alone as symptoms are not necessarily relieved by oxygen. The onset may be delayed six to 96 hours after arrival at higher altitudes. Altitude headache tends to be aggravated by movement, coughing, straining, or exertion. Evidence suggests that this may be due to an increase in intracranial pressure, based on the findings of papilledema, retinal hemorrhages, and elevated cerebrospinal fluid pressure on lumbar puncture. Treatment has included the use of mild analgesics to relieve the pain, and furosemide, acetazolamide, and dexamethasone to relieve intracranial hypertension. Altitude headache may be aggravated by an underlying migraine condition.

Effort/Exertional Headache
Another nonmigranous vascular headache which may be related to environmental or physical factors is the effort or exertional headache. This headache may occur in a variety of situations such as following intense physical exercise, coughing, or straining (during weight lifting or during sexual activity). The cough headache may be due to organic causes, such as intracranial tumors or the Chiari malformation, although the majority of effort headaches are due to benign

conditions. If this headache is persistent or is associated with vomiting, a structural workup and specialty consultation would is indicated.

Effort and exertional headaches have been observed in highly trained athletics and may be indistinguishable from a migraine headache. These individuals may aggravate their condition by becoming dehydrated, developing excessive heat production from muscle activity, or becoming hypoglycemic from sustained activity.

Immersion Headache
During in-water activity, a distinct effort headache, the immersion headache, is seen in susceptible individuals. This water immersion headache commonly occurs after jumping in and swimming underwater. It results in an explosive, throbbing, severe headache that usually occurs while underwater and reaches its maximum upon surfacing or shortly thereafter. Although the character suggests structural causes, such as subarachnoid hemorrhage, this entity is usually benign. Immersion headache is precipitated by a specific situation and represents a variant of the exertional vascular headache. Immersion headaches tend to be recurrent in repeated water activity situations.

Sexual Headache
The sex associated headache occurs under situations of exertion and may result in a sudden excruciating vascular headache. Headaches usually occur at the time of mounting sexual arousal and also may be aggravated by anxiety, including that precipitated by illicit sexual activity with non spousal partners. Although they are usually not recurrent, these headaches may be incapacitating at the time.

Food/Chemical Headache
A variety of food and chemical substrates may precipitate vascular headaches. These substances are implicated in precipitating both migraine and non-migraine headaches. The non-migraine food associated headache is precipitated only by the substance, and is not otherwise characteristic of migraine. In the migraine food associated headache, classic migraine headache occurs in other situations besides those precipitated by food or chemicals. Chemical substances suspected of triggering vascular headaches include tyramine, phenylethylamine, monosodium glutamate (MSG), nitrites, and aspartame. Tyramine and phenylethylamine are in foods such as aged cheese, chocolate, yogurt, buttermilk, nuts, bananas, onions, avocados, figs, and red wines. Nitrites and nitrite compounds are used as food preservatives and to flavor enhancers, and are in foods such as smoked fish, hot dogs, bacon, sausage, baloney, corn beef and pastrami. Monosodium glutamate is a common food additive in oriental dishes, instant and canned soups, potato chip products, processed meat, gravies, TV dinners, and gourmet seasoning. Aspartame, an artificial sweetener used in drinks and other food substances has been implicated in precipitating headaches. Caffeine products have also been implicated in precipitating vascular headaches. Caffeine headaches are associated with excessive caffeine use (coke, coffee, tea, and colas) on weekdays and relative abstinence on weekends. Caffeine has a vasoconstrictive effect on blood vessels, and as it is metabolized and wears off, the blood vessels dilate, which may precipitate a vascular headache.

By judiciously tapering off caffeine or reducing caffeine usage these headaches may be avoided. Medications implicated in vascular headache include reserpine, nitroglycerine, hydralazine, and oral contraceptives. Withdrawal from corticosteroid medication may precipitate headaches. Illicit drugs such as PCP, amphetamines, and cocaine may also trigger vascular headaches.

Migraine Headache Syndromes

Migrainous vascular headaches encompass a number of characteristic syndromes, such as common migraine, classic migraine, cluster headache, and lower half (facial) migraine. Migraine syndromes associated with persistent neurologic deficits include the complicated migraine and acephalgic migraine. Vascular headaches of the migraine type are recurrent attacks of headaches, varying in intensity, frequency, and duration. They are commonly unilateral in onset, associated with anorexia, nausea and vomiting, and preceded by conspicuous sensory, motor, or mood disturbances. These characteristics are not necessarily present in each attack or in each patient. Features suggestive of migraine headaches include childhood onset of headache, cyclic vomiting or carsickness, a lifelong history, strong family history of similar hemicranial throbbing headaches, and response to ergot medication during the acute headache phase. The most characteristic feature of migraine is the classic prodrome or aura that precedes the headache. Symptoms of the migraine aura include nonspecific nausea, vomiting, anorexia, or a variety of transient neurologic symptoms. The incidence of migraine headache in the general population ranges between 3 and 20 percent. In one study of the young adults, 20 percent of males and 50 percent of females described at least one severe headache associated with features of migraine.

Common Migraine   Common migraine accounts for 70 to 80 percent of migraine patients. The prodrome of common migraine tends to be vague and symptoms may last hours rather than the characteristic 20 to 30 minute aura that proceeds classic migraine. The headache is throbbing and usually unilateral, but may be bilateral. A family history of headache occurs in 65 to 90 percent of migraine patients.

Classic Migraine   Classic migraine accounts for 10 to 20 percent of migraine patients. The classic prodrome occurs 20-30 minutes prior to the onset of headache. This prodrome has a characteristic march, that is, the symptoms seem to build in intensity over a 20 to 30 minute period. The migraine accompaniment is the transient neurological event which accompanies the migraine. Neurological manifestations are often contralateral to the side of the headache. Symptoms include visual, sensory, motor, or speech disturbance. Visual symptoms described include visual distortions or flickering lights. The classic fortification scotoma (teichopsia), is described as jagged streaks of light, resembling a sawtooth that shimmers and spreads from the central to the peripheral or from the peripheral to the central vision, over 20 to 30 minutes. Other visual symptoms include a halo phenomenon (objects appear to have halos around them) or a shimmering heat wave appearance similar to heat radiating off hot pavement. Visual symptoms may be 1) monocular in ocular (retinal) migraine, or 2) bilateral (hemianopic visual fields) in the case of occipital (ophthalmic) migraine. Visual distortions may include alterations in color or size (micropsia or macropsia), tilting of the visual environment, multiple visual images (polyopia), or

persistent visual images (allesthesia). Visual symptoms are usually positive phenomenon, that is, they appear as light as opposed to dark phenomenon (absence of vision). The visual field defect may progress until tunnel vision and actual blindness occur. The symptoms march over a 20 to 30 minute period and are followed by a unilateral throbbing headache. Other migraine accompaniments (transient neurological symptoms), include sensory symptoms, such as the cheiro-oral paresthesia, transient hemiparesis, hemiplegia, dysarthria, or aphasia. These neurological symptoms generally proceed the headache; however, they may occur during or after the onset of headache. Photophobia may accompany the visual symptoms.

Cluster Headache   Another characteristic migraine syndrome is the cluster migraine. This accounts for two to five percent of migraine patients and has a strong male predominance (five or six to one, male to female), usually affecting the young adult. Cluster headaches are named because of their seasonal cluster and tendency to occur in the spring and fall. They occur approximately one to three times per day, last about l hour, recur over weeks to months, and are followed by headache free intervals of months to years. There are two categories of cluster headaches. The episodic cluster headaches occur with long refractory headache free periods, while the chronic cluster has remissions (headache free periods) of less than 12 months. Cluster headaches are characteristically very severe and disabling, and are often described as boring, searing, or stabbing. Characteristic associated symptoms include lacrimation, rhinorrhea, nasal stuffiness, and a partial Horner's syndrome (ptosis and miosis). Unlike common or classic migraine, where the patient seeks a quiet room and rest, the cluster patient will pace and walk around. Cluster headaches tend to occur in very perfectionist, obsessive-compulsive people. During the susceptible cluster period, the patient is sensitive to alcohol. Even very small doses of alcohol may precipitate a cluster attack. Only 15 to 30 percent of cluster patients have a family history of headaches, which is less than the usual 50 to 90 percent positive family history in common and classic migraine patients. On initial presentation, the cluster headache will usually be referred for specialty consultation and structural workup to rule out intracranial pathology. Headaches tend to occur during periods of REM sleep and the patient often awakens from sleep with a severe headache. The mainstay of therapy is prophylactic treatment with Sansert (methysergide), lithium, ergotamine, and oxygen therapy. Generally these patients are NPQ for general military duty, due to the severe incapacitation associated with this headache.

Complicated Migraine   Neurological symptoms associated with migraine headaches are called migraine equivalents or accompaniments. Neurological symptoms and signs that persist beyond the headache are called persistent migraine equivalents, and when they last over 24 hours the condition is called complicated migraine. Persistent or complicated migraine equivalent syndromes include the hemiplegic migraine, basilar artery migraine, and ophthalmoplegic migraine. A number of other less common syndromes are the dysphrenic migraine, recurrent migrainous vertigo, abdominal migraine, cardiac migraine, and paroxysmal tachycardia. Hemiplegic migraine consists of persistent hemiparesis or hemiplegia following a headache and generally there is a strong family history. Basilar artery migraine is a migraine which is restricted to the posterior cerebral circulation. It usually presents in childhood and often has a strong family history of headache. The headache is followed or proceeded by symptoms of paresthesia, vertigo,

ataxia, dysarthria, and occasionally, transient loss of consciousness. Ophthalmoplegic migraine is a rare migraine syndrome, usually presenting in early childhood or young adolescence. The patient presents with pain followed by extraocular muscle palsy ipsilateral to the side of the headache. The cranial nerves affected are, in order of decreasing frequency of involvement, the Oculomotor (III), Abducens (VI), Trochlear (IV), and Trigeminal (V). The ophthalmoplegia may persist for several weeks. The pupil may not be spared, with both sympathetic and parasympathetic pupil dysfunction. Generally there is no aura preceding the headache. Because of the persistent neurological deficits, urgent specialty consultation and structural workup is indicated. History of recurrent, complicated migraine in applicants is disqualifying for general duty.

Acephalgic Migraine Approximately 20 percent of migraine patients do not experience headaches with their neurological symptoms. A migraine equivalent not associated with a headache is termed an acephalgic migraine. Migraines associated with neurological signs or symptoms beyond 24 hours, particularly when not associated with a headache, represent a complicated diagnostic challenge. On initial presentation these patients deserve a through workup for vasculitis and other forms of vascular disease such as atherosclerosis, embolic disease, etc. The most common migraine symptoms to occur without headache are visual symptoms. Visual symptoms confined to one eye (transient monocular blindness) may mimic amaurosis fugax due to vascular disease. Characteristic migraine visual phenomenon are bright or flashing phenomenon, while embolic or atheromatous phenomenon affecting the retinal circulation tend to be dark rather than light. Acephalgic migraine symptoms usually spread over 20 to 30 minutes, which is characteristic of migraine, while transient cerebral ischemic attacks (TIA) usually develop suddenly without a march of symptoms. A seizure disorder may march, but the seizure marches over seconds or minutes. Another common neurological symptom to occur without headache is paresthesia (transient sensory phenomenon). Other transient symptoms include speech difficulties (dysarthria), alexia, and recurrent vertigo. In children, acephalgic migraine may be manifested by acute confusional states, transient global amnesia, and dysphrenic states (psychic or mood migraine).

Facial Migraine Facial migraine presents in the older population with jaw, neck (carotidynia), periorbital, or maxillary pain and is described as sharp ice pick like jabs. Serious medical conditions, such as temporal arteritis or cerebral vascular insufficiency, must be differentiated from this condition. Temporal arteritis occurs in patients over 60 years of age who complain of jaw claudication, headache, fatigue, polymyalgia rheumatica, and have an elevated ESR and anemia on laboratory studies. The diagnosis is made by temporal artery biopsy. Temporal arteritis is treated with corticosteroids to prevent symptoms of blindness or neurological dysfunction. Carotidynia or lower half headache is usually diagnosed when the other studies fail to identify either ischemic vascular disease or temporal arteritis.

Migraine Pathogenesis The theory behind migraine has traditionally involved vascular dysfunction. Presumably, the neurological symptoms are due to vasoconstriction and the headache is due to vasodilatation resulting in stretching of pain sensitive fibers in the blood vessels. Other factors appear to be evident as well, however. Other phenomenon associated with

migraine include a spreading depression of cortical activity, preceded by increased metabolic activity, which progresses across the cerebral cortex. Regional blood flow studies have indicated that in classic migraine, hypoperfusion (reduction in blood flow) occurs over the cerebral cortex and spreads at a rate of two to three millimeters per minute. This reduction in cortical flow may be a manifestation of neural dysfunction rather than a primary vascular problem. During the prodromal phase, there is an increase in serotonin release from the platelets, which increases platelet adhesion and aggregation in the blood vessel. This is followed by a decrease in serotonin levels during the headache. Prostaglandins, platelet factor 4, and beta thromboglobulin, may also be increased, resulting in platelet emboli, possibly aggravated by vascular endothelial changes.

Treatment of Headaches
In general, if a precipitating factor can be found that aggravates or causes a headache, reducing or eliminating this factor may reduce or prevent the headache. A careful diet history and avoidance of provocative foods and substances may relieve headaches. Alcohol consumption should be tapered and caffeine history, if considered excessive, should be considered as a possible cause. Hormonal changes seem to be implicated, as migraines are more common in women. As estrogen levels fall, migraines may be precipitated. This accounts for the increase in migraines during the premenstrual period and a change in migraine character with menopause or hormonal manipulation. Pregnancy may also alter the migraine (favorably or unfavorably) and oral contraceptive use is implicated in increasing migraine severity. There is an increased likelihood of an ischemic vascular event in a patient with a history of migraine and oral contraceptive use who smokes.

Psychological factors such as stress, fatigue, and sleep deprivation should be avoided if possible. Physical factors known to precipitate headaches, such as exertion, exposure to smoke, solvents, or glare should be avoided.

Vascular migraine headaches are approached three ways: (1) symptomatic therapy for the infrequent headache, (2) prophylactic therapy if the headache occurs more than once or twice a week or is associated with severe incapacitating pain or neurological symptoms, and (3) abortive therapy if a classic prodromal phase occurs. Ergotamine remains the single most effective abortive agent and is administered either sublingually, intravenously, or rectally. Gastrointestinal motility is reduced during migraine attacks and delays absorption of orally administered medication. Prophylactic therapy includes beta blockers (propranolol), tricyclic antidepressants (amitriptyline or nortriptyline), and calcium channel blockers (nifedipine, diltiazem). Symptomatic therapy includes a variety of analgesic and anti-inflammatory medications.

VIII. MANAGEMENT OF ACUTE SPINAL CORD INJURIES

Introduction
Injury to the spinal column and spinal cord represent one of the most significant injuries in terms

of medical complications and economic impact. Like head trauma, spinal cord injury can be divided into penetrating and nonpenetrating injuries. The majority of the injuries are nonpenetrating, and are usually due to deceleration forces from motor vehicle accidents, airplane crashes, falls, diving accidents, contact sports, or crush injuries. The injury may affect the spinal column (bone and ligaments), spinal cord (neural elements), or both.

Classification of Spinal Injuries
Spinal injuries are classified according to: 1) the mechanism of impact leading to the injury, or 2) the pathophysiological damage to the spinal cord and column. Spinal column injuries are divided into: 1) fractures, 2) dislocations, or 3) fracture-dislocations (the most common type). Following initial stabilization of the injury, it is important to identify the level of injury, bearing in mind that approximately one third of spinal injury patients may have other systemic injuries or another level of the spine affected.

Spinal cord injuries may be classified according to the area of cord damaged or the extent of clinical syndrome: (complete versus partial; anterior cord, posterior cord, or central cord). Vertebral injury may be classified according to the direction of force vectors applied to the vertebral column. These injury patterns include flexion, extension, axial compression, lateral flexion, rotation, or a combination of patterns. Flexion and extension may be further subdivided into disruptive or compressive injury (flexion or extension). Certain predisposing factors can aggravate or precipitate acute spinal injury, such as preexisting spondylitic disease, osteoporosis, ligament hypertrophy, or spinal stenosis.

Evaluation of Spinal Injuries
Spinal injury, particularly cervical spinal injury, should be suspected in anybody who has sustained an injury above the clavicle, or who has sustained a head injury and is unconscious. Other clues to the presence of a spinal cord injury include neck or back pain, tenderness on palpation, a step-off deformity, muscle spasm or swelling, an electrical sensation with neck movement, flaccid extremities, absent reflexes, incontinence, diaphragmatic breathing, or asymmetric weakness (distal greater than proximal), and priapism.

Management of Spinal Injuries
As with management of acute head trauma, the most important aspect is to ensure the ABC's, (airway, breathing, and circulation). Throughout the initial management of the trauma patient it is extremely important to prevent further damage to the spinal cord. This is accomplished by avoiding flexion or extension of the neck, and maintaining neutral head position. Prior to extraction, the patient should be placed on a short spine board and immobilized with sandbags, tape, or straps. Plastic IV bags may be used in lieu of sandbags. The patient should be lifted onto a long spine board and then secured with straps. Obviously in a situation where the patient is in a dangerous position, such as a burning aircraft, these considerations would have to be hastened or bypassed to save the individual's life. In general, the plastic Philadelphia collar or the Hare extrication collar should be used in combination with sandbags, tape, and spine boards. These collars primarily limit extension, but do little to limit flexion.

Examination should include palpation of the cervical, thoracic, and lumbar spine, an adequate motor, sensory, and reflex examination of the upper and lower extremities, and a rectal exam. Continued reassessment of the ABCs and neurological examination are indicated. Significant injuries of the upper thoracic spinal column are often associated with respiratory distress from flail chest, hemopneumothorax, or circulatory compromise from aortic arch dissection, myocardial contusion, or cardiac tamponade. Injuries to the lower thoracic spinal column are often associated with intra-abdominal injury and renal damage. Delayed neurological deterioration in a spinal injury patient could signify the development of a spinal epidural hematoma, spinal abscess, or vascular or neural compromise of the spinal cord. Injuries to the thoracic and lumbar spinal column are common complications of aircraft accidents, and occur in 30 to 60 percent of ejections or crash landings. In acceleration/deceleration in the G plane, the greater mobility of the cervical spinal column accounts for a higher incidence of injuries to the cervical spinal cord. Injuries in the thoracolumbar area may result in significant neurological sequela, because there is less space available for the cord in this region. Spinal cord blood supply in the thoracolumbar region is tenuous compared to the high thoracic and cervical areas. Injury forces required to injure the thoracic spinal column involve a greater amount of destruction and displacement, which may result in intra-thoracic and intra-abdominal injuries.

Prior to transport of the spinal injury patient, it is extremely important to adequately pad areas that have become anesthetic from the spinal injury. Casting of an anesthetic extremity should be avoided. Alternatives include bivalved casts, splints, or external fixation devices. Spinal cord injury patients often have urinary drainage complications and may require intermittent catheterization or an external condom catheter. Treatment with ascorbic acid and Mandelamine helps to reduce urinary tract infections. Prophylactic treatment with antacids is also important as these patients are prone to stress ulcers. Long term complications from spinal cord injury include pneumonia, pulmonary embolism, gastrointestinal hemorrhage from ulcers, renal stones, urinary tract infections, and decubitus ulcers. Attention to the nursing management problems in spinal cord injury patients is essential to preclude or alleviate these complications. The use of glucocorticoids and antibiotics remains a controversial area and should be given only at the direction of specialty consultants. Referral to a neurosurgical center should be accomplished as soon as feasible for any patient with a neurological deficit or unstable spinal injury.

Spine Radiography
Following spinal stabilization on a long spine board and a neurological evaluation including sensory, motor and reflex examination, the patient should undergo radiographic evaluation (see cervical spine radiology sheet). The acutely injured patient should undergo cross table lateral C-spine X-ray which should include the C7-T1 level. When stable, AP radiographs of the cervical, thoracic, and lumbar spine should be obtained, as well as an open mouth odontoid view. Radiographic findings that may simulate fractures or ligament injuries include the pseudosubluxation of C2-C3 (seen in one to seven year olds), incomplete ossification of the posterior elements, spina bifida, the mach band variant, unfused secondary ossification centers (apophysis), butterfly vertebra, or soft tissue ossification.

Evaluation of cervical spine films
  Look for the following criteria:
  • Soft Tissue
    ▪ Widened retropharyngeal space (C-2) > 7 MM
    ▪ Widened prevertebral fat stripe (C-3) > 5 MM
    ▪ Widened retrotracheal space (C-6): child > 14 mm, adult > 22 mm
    ▪ Tracheal deviation/laryngeal dislocation on AP film.
  • Vertebral Alignment
    ▪ Loss of lordosis
    ▪ Kyphotic angulation >11°
    ▪ Torticollis
    ▪ Widened interspinous space
    ▪ Vertebral body rotation
    ▪ Space available for cord is <14mm.
  • Joint Abnormalities
    ▪ Axis-Dens interspace (ADI): child (< 8 yr) > 5 mm, adult > 3 mm
    ▪ Disc space disruption
    ▪ Apophyseal joint widening.

Use of the Gardner-Well Tongs
Ideal management of the suspected spine injury patient involves the use of skeletal traction, such as the Gardner-Wells or Crutchfield tongs. These require either mechanical weights or spring tension devices, which apply a force of usually seven to 10 pounds. In aeromedical evacuation, the William's traction apparatus, which is attached to the standard military litter, is the preferred method of applying traction to the tongs. The Williams traction apparatus provides tension from a spring device and avoids the swinging weights which might aggravate a medevaced spinal cord injury patient.

The Gardner-Wells tongs should be placed over the patient's scalp. If time permits, an area of scalp over the tong insertion point should be shaved and prepped with an antiseptic solution and infiltrated with local anesthetic. Place the Gardner-Wells tongs approximately two finger widths above the external ear in the plane of the external auditory canal. The squamosal line, where the temporalis muscle inserts, is a helpful landmark. Tong placement should be below this line to allow adequate traction. The tongs are screwed in equally on both sides and a small spring-loaded protuberance will stick out of one side of the tongs when adequate tension is applied. The securing nuts should be tightened to prevent the tongs from loosening. The tongs should be readjusted one day later, again setting the tension spring so that it sticks out approximately one millimeter from the spring-measuring device. This is the only time that the tongs should be readjusted, as further tightening will result in erosion of the tong point through the skull with obvious complications. Patients with skeletal traction should have lateral C-spine views daily, and weights are changed, to assess vertebral alignment.

Back Pain

Low back pain is one of the most common conditions affecting Americans. It costs an estimated $16 billion a year in lost wages and medical costs, and disables approximately five and one half million Americans. It is estimated that the lifetime prevalence of low back pain is 40 percent in the U.S. Approximately one percent of these will also have localizing extremity symptoms (radiculopathy or sciatica). Low back pain is clearly an occupational disease, and is associated with activities requiring heavy lifting and exposure to vibration. Back pain can be divided into four phases: acute, subacute, chronic, and recurrent. Acute low back pain resolves within six weeks after onset and accounts for 75 percent of the of back pain population. Only 20 percent of these patients will have a clearly identifiable diagnosis. Subacute back pain resolves within 12 weeks, and accounts for about 10 percent of all back pain patients. Chronic low back pain lasts over 12 weeks and accounts for 5 percent of the low back population. Recurrent low back pain, often a disabling condition, accounts for approximately 10 percent of the low back pain patients. Chronic and recurrent back pain patients account for 85 percent of the low back pain costs.

Management of Acute Low Back Pain
As with any neurological complaint, the most important initial goal is to establish whether or not a life- threatening condition exists. In the management of acute low back pain, several factors may suggest a possible early presentation of a serious condition. Urgent evaluation should be considered for any patient who is in severe, writhing pain, as this may be the early presentation of an intra- abdominal vascular process, such as a dissecting abdominal aortic aneurysm. Patients who have significant pain at rest may be harboring an infectious or neoplastic process involving the spinal cord or column. Finally, any patient with an evolving neurological deficit, such as sacral anesthesia, bowel or bladder incontinence, or progressive sensory motor dysfunction, should be referred to an appropriate center for urgent evaluation.

The mainstay of treatment for acute low back pain is bedrest. Recent studies have shown that two days are as effective as seven days of bedrest and result in 45 percent less time lost from work. Generally in a military environment, where a patient is either fit or not fit, it is often not feasible to return a patient to partial work status, i.e., prolonged bedrest may be indicated in certain military occupational rates. During the bedrest phase, a variety of medications can be considered, such as analgesics, muscle relaxants, or nonsteroidal anti-inflammatory medication. Drugs with a high narcotic potential, such as Percocet or Percodan, should be avoided and Valium should not be used as a muscle relaxant as it also has a serious side effect of depression. In some situations, tricyclic antidepressants are effective as analgesics.

Upon resolution of the severe back pain when the patient is ambulatory, a variety of physical therapy programs should be considered, including strengthening exercises, range of motion, ultrasound, heat and cold packs, and transcutaneous nerve stimulation. In general, gravity traction or bedrest traction is ineffective and can lead to serious secondary complications and should be avoided. One of the most important aspects following resolution of acute low back pain is the back education program, or "low back school", available in some physical therapy departments. An evaluation by an experienced physician of the workplace may lead to improvements in occupational procedures to prevent further recurrences of low back pain.

Manipulation may temporarily decrease pain but has no long-lasting benefit. In general, manipulation with rapid changes of direction may actually further weaken spinal ligaments. Soft tissue massage and pressure point techniques may be better tolerated.

Conservative therapy of acute low back pain with sciatica is usually effective, as 50 percent of patients with sciatica will usually resolve their symptoms within six weeks. Those who fail to respond to conservative therapy should be referred for surgical intervention. Patients whose symptoms continue for more than six weeks should undergo further medical evaluation including a complete blood count and sedimentation rate, and be considered for radionuclide bone scan and lumbosacral spine X- rays.

Lumbar Radiography Indications for spinal X-rays include age over fifty years, history of trauma, history of cancer, unexplained weight loss, pain at rest, illicit IV drug use, steroid use, fever, neurological deficit, and medicolegal considerations. In most cases spinal radiographs are normal or show only nonspecific findings. However, there may be findings on the X-ray which suggest certain etiologies of back pain, such as:

> Spondylolysis
> Spondylolisthesis
> Disc narrowing
> Schmorl's nodes
> Lumbarization of S-1
> Sacralization of L-5
> Osteophyte formation
> Traction spurs
> Facet sclerosis
> Vertebral sclerosis

Sciatica Sciatica, or lumbar radiculopathy, is manifested by pain, weakness, or sensory loss in a nerve root distribution in the lower extremity. Although it is commonly due to a herniation of the nucleus pulposus with impingement of the nerve root, it may also be caused by compression of the cauda equina from tumor, abscess, or hemorrhage. It may also be due to impingement of the nerve root by hypertrophy of the lumbar facets, causing spinal stenosis. Other less common causes include congenital anomalies of the nerve roots, nerve and bone tumors, metastatic disease, and degenerative synovial cysts (Tarlov cysts). Sciatic leg pain may also be caused by extraspinal involvement of the lumbosacral plexus, by tumors or endometriosis involving the pelvic peritoneum, by compression of the sciatic nerve near the hip due to external compression from a wallet or prolonged sitting, or by localized tumors of the sciatic nerve.

Despite this rather extensive differential of sciatica, the majority of cases are related to a degenerative condition of the lumbar disk. The most likely levels involved are the L4-L5 disk causing an L-5 radiculopathy, or the L5- S1 disk causing an S-1 radiculopathy. The L-5 radiculopathy causes weakness of the dorsiflexors and evertors of the foot, and numbness and

pain over the lateral aspect of the leg and ankle and dorsal aspect of the foot. The S-1 radiculopathy results in weakness of the ankle plantar flexors and hamstrings and numbness and pain over the lateral aspect of the sole of the foot.

Another clinical entity is lumbar neurogenic claudication, usually due to lumbar spinal stenosis. Narrowing of the central canal and lateral aspect of the spinal column results in low back pain and bilateral leg pain primarily while ambulating. This condition often mimics vascular insufficiency of the lower extremities. Lumbar neurogenic claudication, seen with degenerative spine disease, is characterized by the lack of signs of vascular insufficiency (atrophic skin and diminished distal pulses). Neurogenic pain usually resolves after resting for 15 or 30 minutes, whereas pain due to vascular insufficiency, which is usually confined to the calves, resolves immediately with rest. Persons with lumbar spinal stenosis walk in the flexed position to avoid the prominent pain and weakness caused by central canal compression.

Evaluation of the Lumbar Spine and Lower Extremity Nerve Roots Examination of the lumbar spine should include evaluation of the spinous processes and their alignment. Look for excessive lordosis, scoliosis, and vascular skin lesions (birthmarks). During examination of the extremities, assess any muscle atrophy. Include spinal range of motion (extension, flexion, lateral flexion, and lateral rotation). Extreme range of motion can be ascertained by having the patient bend and touch his toes. During examination of the muscle groups of the lower extremity, include individual muscle group testing of the hip flexors, extensors, abductors, and adductors; knee flexors and extensors; ankle dorsiflexors, plantar flexors, invertors and evertors; and toe dorsiflexors and plantar flexors. Muscle strength may also be tested by having the patient heel walk, toe walk, hop on one foot, duck walk, and do one-legged deep knee bends.

Provocative maneuvers may detect lumbar disc disease or joint pathology. The straight-leg raising maneuver is conducted with the patient lying supine. The leg is slowly elevated. If pain is reproduced in the back or leg, the angle is noted. Leseque's maneuver is a modification of the straight leg raising sign. The leg is raised to a level just prior to eliciting pain, then the foot is dorsiflexed, and leg or back symptoms are noted. Both are signs of lumbar disc disease. The femoral stretch maneuver starts with the patient in the prone position and the leg extended at the knee. The hip is gradually extended posteriorly. This stretches the lumbar L-4 nerve root. Reproduction of symptoms may be indicative of lumbar disk disease at the L3-L4 level. A maneuver to detect musculoskeletal problems at the hip is Patrick's sign or the FABERE maneuver, which stands for hip Flexion, Abduction, External Rotation and Extension.

Sensory examination of the lower extremities includes light touch and pinprick. If bowel or bladder symptoms are present, sensation around the anus and perineal region should also be tested. Reflex examination should include the quadriceps (knee jerk) and gastrocnemius (ankle jerk) reflex. The cremasteric and bulbocavernosus reflex should be tested if the patient has bowel or bladder symptoms. Patients who fail to respond to conservative therapy and have signs of radicular symptoms over six weeks should be referred for neurosurgical or orthopedic evaluation. Patients with low back pain whose symptoms are unremitting or severe, or who have profound

weakness should be evaluated on an urgent basis, particularly if there indications of a neoplastic or infectious process. Chronic low back pain may occur in a variety of hereditary and metabolic conditions, such as spondylolysis, osteochondrosis (Scheuermann's disease), osteoporosis, ankylosing spondylitis, fibromyalgia, idiopathic sclerosis, Paget's disease, and vertebral body fusion (Klippel-Feil syndrome).

Neck Pain and Upper Extremity Radiculopathy
A variety of conditions may cause pain in the neck or upper extremities. Perhaps the most common is cervical spondylosis or disc disease of the cervical region. The most common disc syndrome in the cervical region is a C-6 radiculopathy, which causes weakness of the proximal upper extremity (deltoid, biceps, and wrist flexors), diminished biceps and brachioradialis reflexes, numbness over the thumb and index finger, and pain in the arm radiating to the thumb and index finger. The next most common disc syndrome is a C-7 radiculopathy, which causes weakness of the triceps and wrist extensors, numbness of the middle finger and diminished triceps reflex. The C-8 radiculopathy causes pain in the arm radiating to the ring and little finger and weakness of the hand intrinsic muscles, primarily finger flexors. Cervical disc disease is managed similarly to lumbar disc disease, with bedrest and analgesics as necessary, and physical therapy after the acute phase.

Peripheral Neuropathies
Peripheral neuropathies are due to a variety of etiologies, but in the young active-duty military population, they are most commonly due to trauma or chronic entrapment syndromes. Peripheral nerves may be injured by a variety of physical means, including percussion, traction, compression, ischemia, cold, or transection. In the older age groups, diabetes and alcohol are possibilities, as well as inflammatory peripheral neuropathies. Toxic neuropathies may occur from exposure to a variety of solvents and chemicals used by aviation maintenance and ordnance personnel. Hereditary neuropathies are quite common, and may be cumulative with the effects of other neuropathies.

An injury classification of peripheral nerve injuries is based on anatomic damage. The most common peripheral nerve injury is neuropraxia, which is a localized (segmental) demyelination. This type of nerve injury will usually resolve within hours to days. The next injury type is axonotmesis, which is damage to the axon cylinders of the nerve. Damage of this type requires a longer period for recovery and may take months for recovery. The last, and worst, type of injury is neurontomesis, which is a disruption of both the axon cylinder and myelin. It is commonly due to laceration, with the nerve ends no longer in physical continuity with each other.

Entrapment Neuropathy Syndromes
Suprascapular Neuropathy This neuropathy is due to entrapment of the suprascapular nerve at the shoulder. It is also called rucksack palsy, from the straps of a heavy rucksack compressing the suprascapular nerve. This is a pure motor nerve disorder and causes weakness of external rotators of the arm and the shoulder abductors.

Median Nerve Entrapment Neuropathies.

Pronator Teres Syndrome   The median nerve may become entrapped at several locations. Entrapment of the median nerve may occur in the antecubital fossa (medial elbow), where the median nerve passes between the pronator teres muscle. The pronator teres syndrome may be seen in a variety of conditions. It may affect weight lifters who overdevelop their forearm muscles, or in pilots who roll their flight suits over the forearms. It causes weakness of the wrist and finger flexors. Entrapment of the proximal median nerve causes weakness of the flexors and of the first and second fingers and thumb. This is called the papal hand sign, because of the characteristic appearance of the hand on attempting to make a fist.

Anterior Interosseous Nerve Syndrome   A branch of the median nerve may become entrapped in the lateral forearm. The anterior interosseous nerve syndrome, or honeymoon palsy, may occur when a spouse's head rests on the forearm overnight, resulting in weakness of the thumb and index finger flexors and pronator quadratus. This results in difficulty with pinching movements of the thumb and index fingers, and pronation of the wrist.

Carpal Tunnel Syndrome   The most common entrapment neuropathy, the carpal tunnel syndrome, results from entrapment of the distal median nerve in the wrist as it passes through the carpal tunnel. This causes weakness of the LOAF muscles (lumbricals, opponens, abductor and flexor of the thumb), which results in atrophy of the thenar eminence. Because of weakness of the thumb muscles, the thumb falls back into the plane of the hand, resulting in the "simian hand" or monkey hand. Tapping the median nerve over the wrist may cause an electrical sensation, characteristic of nerve entrapment (Tinel's sign).   Symptoms may also be reproduced by wrist hyperflexion (Phalen's sign), further compressing the nerve.

Ulnar Entrapment Neuropathies   The ulnar nerve may be entrapped above the elbow by the ligament of Struthers, at the elbow in the olecranon groove, and below the elbow in the cubital tunnel. This causes a sensory loss of the little finger and the lateral aspect of the ring finger, and weakness and atrophy of the hypothenar eminence, resulting in the "claw hand" deformity. Percussing the nerve above, at, or below the elbow may cause electrical sensations or pain, which is diagnostic of entrapment at that region (Tinel's sign). Entrapment of the ulnar nerve at the elbow is very common with trauma, particularly fractures of the elbow, athletic injuries, or chronic compression over the ulnar groove from pressure to the elbow. The ulnar nerve may also be entrapped in the wrist in Guyon's canal. The distal ulnar nerve is a pure motor nerve; entrapment results in weakness of the hypothenar muscles as well as weakness and atrophy of the thumb dorsal interosseous muscle, the large muscle between the thumb and index finger over the back of the hand. Weakness of thumb abduction against the index finger is called of Froment's sign. Injury to the wrist, from fractures or chronic pressure (use of power tools), may result in median (more commonly) or ulnar nerve compression at the wrist.

Radial Nerve Entrapment Neuropathies   The radial nerve may undergo damage in the arm at the spiral groove of the humerus, causing paralysis of the wrist extensors, but sparing the triceps. This syndrome is referred to as Saturday night palsy, crutch palsy, or honeymoon palsy, and is due

to pressure over the spiral groove compressing the radial nerve and causing subsequent weakness. This may occur from falling asleep with the arm draped over a chair following a heavy night of partying, having an incorrectly fitted crutch put pressure on the humerus rather than in the axilla, or from having one's spouse fall asleep with the head pressing against the humerus.

Handcuff Palsy The sensory radial nerve can be compressed over the dorsal aspect of the wrist, in the region of the anatomic snuff box. This causes a pure sensory syndrome with numbness over the dorsal aspect of the thumb. Sensory radial palsy is also called handcuff palsy, because it occurs when handcuffs are applied too tightly.

Thoracic Outlet Syndrome Another condition affecting the upper extremity is the thoracic outlet syndrome. It is a neuro-vascular syndrome, due to compression of the lower brachial plexus or blood vessels in the chest. It may result in motor and sensory symptoms in the hand. Provocative maneuvers that reproduce the thoracic outlet syndrome include the costoclavicular maneuver (pulling the shoulders back) and Adson's sign (abducting, and externally rotating the arms over the head, then changing head position). Distal radial pulses should be palpated to assess for a vascular insufficiency problem.

Lateral Femoral Cutaneous Neuropathy The lateral femoral cutaneous nerve entrapment (meralgia paresthetica) is due to nerve entrapment in the inguinal ligament. This nerve innervates the lateral aspect of the thigh and may cause numbness or burning over the lateral thigh. It may be seen in obese people, diabetics, and also may result from tight-fitting flight gear or a heavy utility belt.

Sciatic Nerve Entrapment Sciatic nerve entrapment, called "wallet sciatica" or "toilet seat neuritis", occurs following prolonged sitting, particularly on hard surfaces where the sciatic nerve is compressed, resulting in weakness of the plantar and dorsiflexors and numbness of the entire foot.

Peroneal Neuropathy The peroneal nerve may be entrapped at the fibular head. It may follow acute trauma or prolonged leg crossing, and results in weakness of the dorsiflexors of the ankle and toes, foot evertors, and numbness of the dorsal aspect of the foot. This syndrome may be distinguished clinically from an L-5 radiculopathy, because there is no back pain.

Tarsal Tunnel Syndrome A branch of the posterior tibial nerve may be compressed in the tarsal tunnel in the foot due to tight-fitting boots or prolonged running, resulting in foot pain and weakness of the toe flexors.

Morton's Neuroma Claw foot or Morton's neuralgia may occur with compression of the digital nerves of the toes. It may be quite painful, and may respond to padding placed between the toes.