CHAPTER 2
An Overview of Localization of Function, and Neurological Diagnosis

In considering the patient with neurological disease, accurate diagnosis is required for a determination of appropriate therapy and for the establishment of prognosis. Diagnosis in neurology seeks to answer two essential questions.

1. Where is the disease (lesion) located?

a) Central nervous system versus

b) Peripheral nervous system versus

c) Neuromuscular junction or muscle.

2. What is the nature of the pathology?

PART I: LOCATION AND PATHOLOGY

A. LOCATION OF DISEASE PROCESS: SYMPTOMS AND SIGNS

Patients with neurological disease come to medical attention because of certain symptoms or complaints elicited in the history and certain signs elicited on examination. These symptoms and signs fall into several categories:

1. Disturbance of mental status (cognitive function) and language function related to disease of cerebral cortex.
2. Disturbance of cranial nerve function due to direct involvement of cranial nerves or of brain stem.
3. Disturbance of motor strength reflecting -

a) Disease of the lower motor neuron and the motor unit at the level of the anterior horn cell, or the nerve root or the peripheral nerve or the neuromuscular junction or muscle producing atrophy of the muscle, flaccid weakness and a variable loss of reflex activity or

b) Severe upper neuron disease due to involvement of motor cerebral cortex, or due to the involvement of corticospinal tracts at the level of internal capsule cerebral peduncles, basilar pons, medullary pyramids or lateral columns of spinal cord producing spastic paralysis, increased deep tendon reflexes and a release of the sign of Babinski.

4. Disturbance of motor control and coordination reflecting -

a) Disease of motor cortex and corticospinal tracts: loss of speed and accuracy.

b) Disease of the premotor and supplementary motor cortex / motor association cortex resulting in defects in patterns of movement, defects in the visual and tactile control of movement. Apraxia occurs and the release of automatisms may occur.

c) Disease of cerebellum - resulting in defects in coordination of limb and axial movements and of "balance"

d) Disease of basal ganglia resulting in

1) Slowness of movement (akinesia and bradykinesia)

And/or 2) resting tremor

And/or 3) other dyskinesias

And/or 4) loss of righting reflexes and "balance"

And/or 5) alterations in tone

5. Disturbance of sensation:

a) Primary modalities (pain, touch, vibration) due to disease of peripheral nerve,nerve root, spinal cord or brain stem or thalamus

b) Discriminative modalities (position, sense, graphesthesia, stereognosis, tactile localization, awareness of double simultaneous stimulation) due to disease of posterior column system or cerebral cortex.

6. Pain including headache - the localization of pain may reflect disease of peripheral or cranial nerve, nerve root, spinal cord, brain stem, thalamus, or cortex.

Meninges and blood vessels about the head (intra and extracranial) or increased intracranial pressure, or the sinuses or orbits, or dental system, may provide the origin of headache.

B. PRELIMINARY DIFFERENTIATIONS OF LESION LOCATION:

1. Disease of muscle

E. M. Marcus et al., *Integrated Neuroscience*

a) Patients manifest lower motor neuron disease affecting limbs, trunks and in some cases, cranial nerves. In early cases, however, deep tendon reflexes are preserved

b) Involvement is more often proximal than distal.

c) No sensory symptoms or signs are present.

d) No long tract motor or sensory involvement is present.

e) Cognitive function - mental status is normal but may be affected in certain hereditary types.

f) The major disorders are of two types

1) Hereditary (muscular dystrophy or congenital myopathies)

2) Acquired (polymyositis or dermatomyositis).

2. Disease of neuromuscular junction

a) Patients manifest intermittent dysfunction of lower motor neuron.

b) Involvement of cranial nerve, motor function is prominent, (extraocular muscles and bulbar motor neurons).

c) If limbs are involved - proximal weakness is often greater than distal weakness.

d) Deep tendon stretch reflexes are preserved.

e) No sensory symptoms are present.

f) No long tract findings are present.

g) Cognitive function is not involved.

h) The major disorder is myasthenia gravis, an autoimmune disorder in which antibodies are produced which block and damage the acetylcholine receptor.

3. Disease of peripheral nerve

a) Motor and/or sensory and/or autonomic function are involved.

b) The pattern of involvement indicates disease of:

1) A specific peripheral nerve (mononeuropathy), e.g., median, ulnar, radial, femoral, sciatic, peroneal.

Or 2) a plexus, such as brachial, lumbar sacral (mononeuropathy)

Or 3) multiple peripheral nerves (mononeuropathy multiplex)

Or 4) generalized type- polyneuropathy. This is the most common variety.

c) When motor function is involved the pattern is that of a lower motor neuron type lesion with atrophy, flaccid weakness and loss of reflex function.

d) When sensory involvement is present; some or all modalities may be involved. In the generalized type, the sensory pattern may be a glove and stocking type

e) Cranial nerves may be involved but without direct brain stem involvement.

f) Long tract finding are not present

g) Cognitive function is intact.

h) If compression (entrapment) is present, localized pain may be a prominent feature.

4. Nerve root or spinal nerve disease

a) Motor and/or sensory function may be involved.

b) Pain is often a prominent feature.

c) The symptoms and signs (pain, sensory, motor and reflex) follow a segmental (dermatomal) distribution. Refer to Tables 2-1, 2-2, 2-3, 2-4.

d) When motor function is involved; the pattern is that of a lower motor neuron type lesion as above.

e) Some or all sensory modalities are involved, in a dermatomal pattern. *Fig. 2-1.*

f) Cranial nerves are not involved.

g) There are no long tract findings - unless, spinal cord is also involved.

h) Cognitive function is not involved.

Dermatomal-Radicular Patterns

Fig 2-2 compares dermatomal and peripheral nerve patterns.

5. Spinal Cord:

a) Lesions are either

1) Transverse (segmental)

2) System involving a system of neurons or fibers over many segments

3) Multifocal producing spotty lesions over multiple levels of the CNS

Transverse lesions

a) Transverse lesions are usually extrinsic with the implication of compressive mass and

possible surgical therapy.

b) Four exceptions are intrinsic lesions, which involve multiple adjacent segments: syringomyelia intrinsic spinal cord tumors (astrocytomas and ependymomas), infarcts due to occlusion of the inferior spinal artery and transverse myelitis.

c) Generally both motor and sensory function are involved.

d) When the motor long fiber systems are involved, the signs of an upper motor neuron lesion are produced: spastic weakness or paralysis, increased deep tendon reflexes and the sign of Babinski.

e) Involvement of the long fiber sensory systems may produce involvement of all modalities below the lesion in a transverse lesion or selective involvement of specific sensory modalities, e.g., position and vibratory sensation when the posterior columns are involved and pain and temperature when the lateral spinothalamic tract is involved. Syringomyelia involves the decussating pain and temperature fibers in the anterior white commissure of the cervical area producing a selective dissociated pain and temperature sensory loss over the shoulders and arms "cape like".

f) Transverse lesions producing damage to the anterior horn cell or anterior root will produce signs of a lower motor neuron lesion at the segmental level of involvement. Fasciculations, twitching of muscle, will be noted in the Segment involved. Fasciculations are indicative of disease of anterior horn cells.

g) Transverse lesions producing damage to the posterior root will produce a local segmental defect at the level of involvement.

h) Cranial nerves are not involved.

i) Mental status, cognitive function, is not involved.

TABLE 2-1: SEGMENTAL SENSORY PATTERNS (FIG. 2-1 AND 2-2. SEE ALSO 8-18 FOR THE UPPER EXTREMITY DETAILS)

The following radicular patterns should be compared to peripheral nerve patterns	
C2	posterior scalp versus trigeminal
C6, C7, C8	the hand
a. C6	thumb and index finger
b. C7	middle finger
c. C8	ring and little (5th) finger
T1	axilla
T5, T6	xiphoid process
T10	umbilicus
T12	above inguinal ligament
L4	medial calf, patella and lateral thigh
L5	lateral calf and medial foot
S1	posterior calf and lateral foot
S2	posterior thigh
S3, 4, 5-	perianal areas.

Concept of Level of Lesion at the Spinal Cord Level

In transverse lesions it is then evident that several determinants of level are considered.

1. Local anterior root or anterior horn level indicated by segmental atrophy and segmental flaccid weakness.

2. Local segmental sensory level indicated by segmental (radicular) loss of all modalities of sensation.

3. Local segmental losses or depression of deep tendon reflexes due to loss of the afferent or efferent component of the monosynaptic stretch reflex.

4. Segmental long motor tract level with presence of spastic weakness, increased deep tendon reflexes and release of the sign of Babinski below this level.

5. Segmental long sensory tract levels indicated by posterior column (uncrossed) and lateral spinothalamic (crossed) deficits below this level.

TABLE 2-2: MOTOR - RADICULAR MUSCLE INNERVATIONS

1. Shoulder muscles,	C4, C5, C6
2. Biceps	C5, C6
3. Triceps	C6, **C7**, C8
4. Intrinsic hand muscles	C7, C8, T1
5. Hip flexors, iliopsoas	L2, **L3**, L4
6. Quadriceps	L2, **L3**, L4
7. Gastrocnemius	LS, L5, **S1**, S2
8. Dorsiflexors of foot (peroneal)	L4, L5, S1

* Bold indicates major innervations

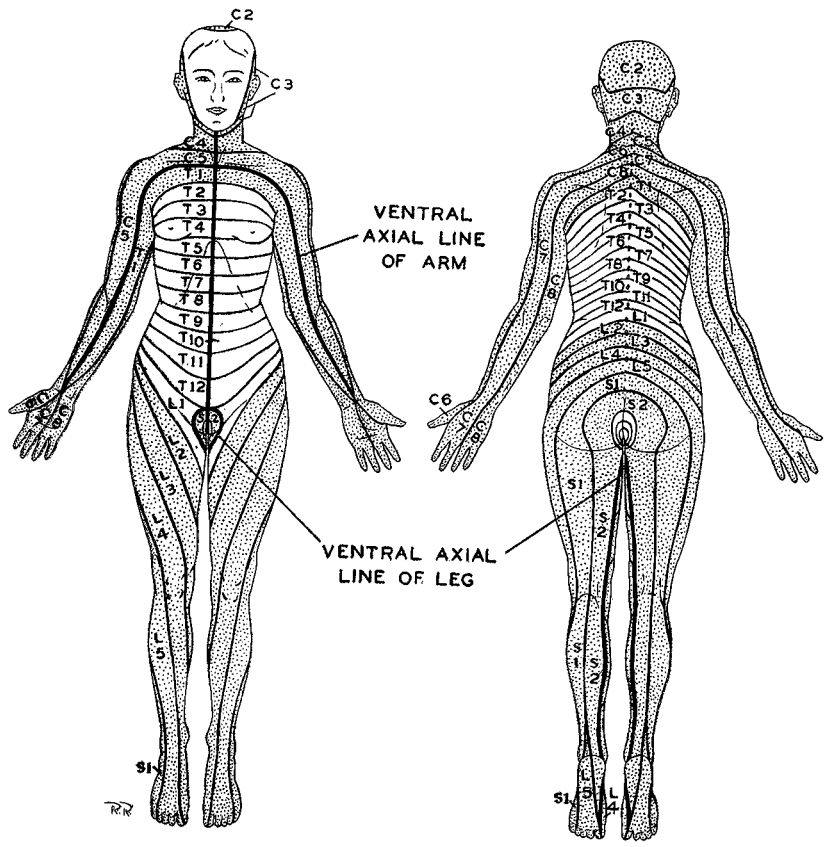


Figure 2-1 Dermatome charts of the human body determined by the pattern of hypalgesia following rupture of an intervertebral disk. From Keegan, J.J., and Garrett, F.D.: Anat. Rec., 102:411, and 1948 (Wiley)

TABLE 2-3: DEEP TENDON STRETCH REFLEXES

1. Jaw	cranial nerve V - pons
2. Biceps	C5, C6
3. Triceps	C6, **C7**, C8
4. Brachioradialis (Radial periosteal)	C5, C6, C7
5. Fingers -	C7, C8, T1
6. Patellar-	L2, **L3, L4**
7. Achilles	L4, L5, **S1**, S2

* Bold indicates major innervations

TABLE 2-4: SUPERFICIAL REFLEXES

1. Upper abdomen -	T7, T8, T9, T10
2. Lower abdomen -	T10, T11, T12
3. Plantar	S1, S2

6. Segmental autonomic level deficits in autonomic function below this level.

6. Brain Stem

a) Cranial nerves findings are present with the specific lower motor neuron or sensory deficits dependent on the level of involvement.

1) CN 12, 11, 10, 9 due to medullary lesions

2) CN 8, 7 and 6 due to pontomedullary involvement

3) Trigeminal - 5 midpontine

4) CN 3 and 4- midbrain

b) Selective involvement of pain in the trigeminal distribution may be present due to involvement of the descending spinal tract and nucleus of the 5th nerve.

c) Long tract motor findings may be present which are unilateral or more often bilateral.

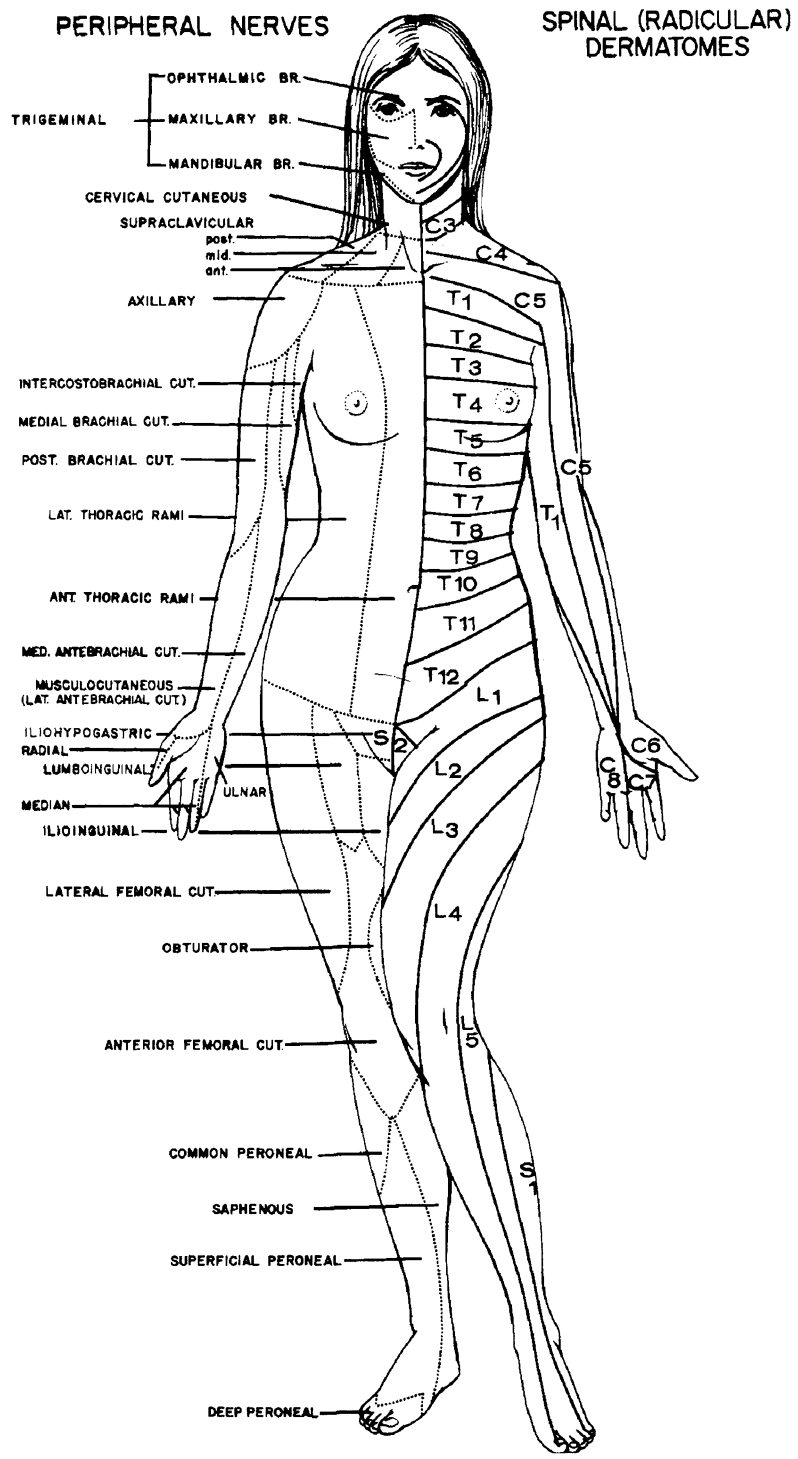


Figure 2-2 Comparison of radicular (dermatome or segmental) and peripheral nerve innervation. A.) anterior

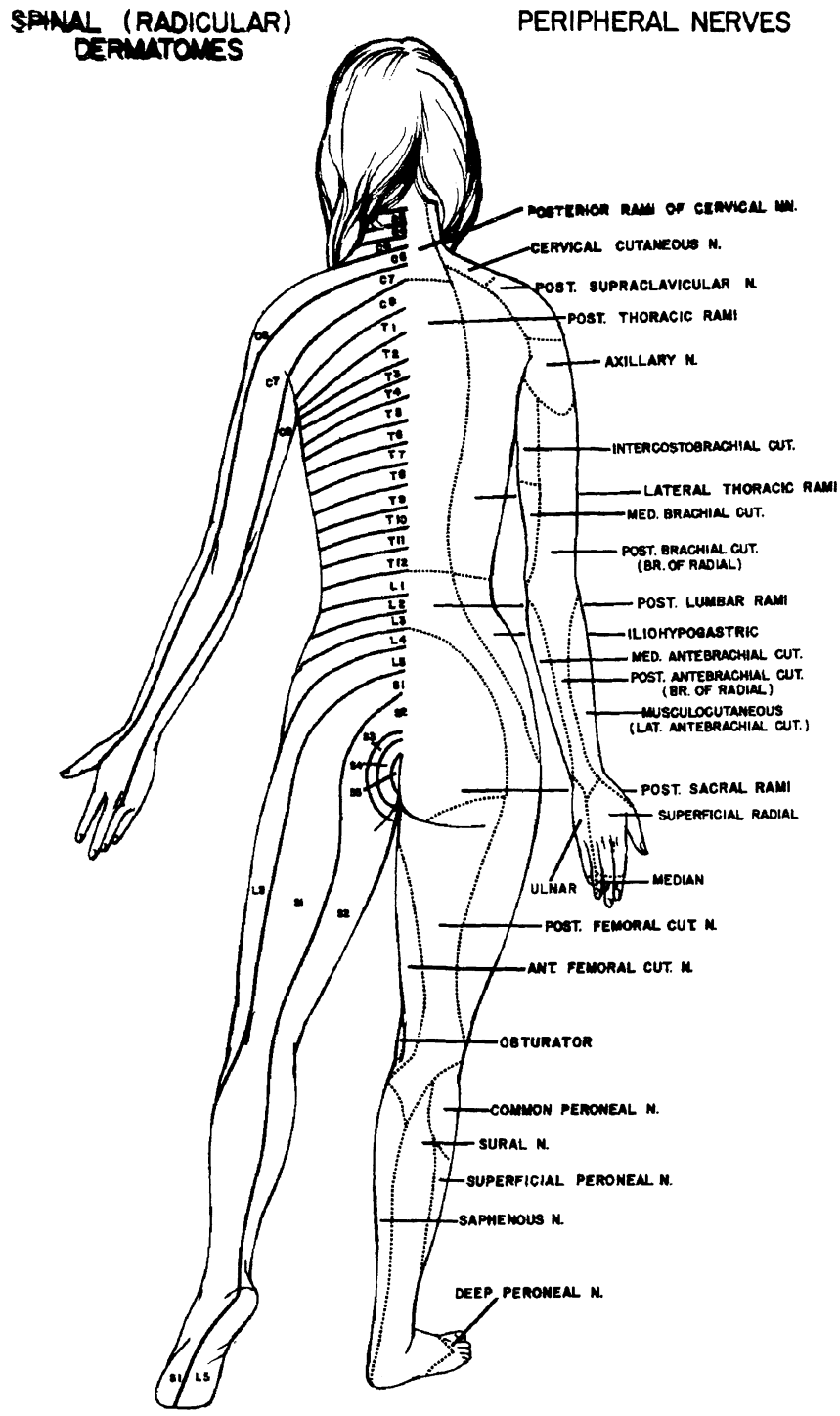


Figure 2-2 Comparison of radicular (dermatome or segmental) and peripheral nerve innervation. B.) posterior view

d) Long tract sensory findings may be present which may be selective, that is dissociated e.g., pain and temperature only when unilateral or bilateral and nonselective. Medial lemniscus or lateral spinothalamic may be involved.

e) Cerebellar pathways may be involved producing alteration in balance, stance, gait and appendicular coordination.

f) In general, mental status, cognitive function is well preserved although level of consciousness may be depressed.

g) Particular combinations of cranial nerve finding ± long tract findings are diagnostic.

1) The jugular foramen syndrome (cranial nerve 9, 10, 11).

2) The cerebellar pontine angle syndrome: (cranial nerves 7 (facial) 8 (auditory and vestibular) and cerebellum plus or minus CN 5, 9, 10, 11.

3) The lateral medullary syndrome (lateral medullary tegmentum) "Wallenberg's syndrome", or syndrome of the posterior inferior cerebellar artery.

4) Ipsilateral peripheral facial weakness and contralateral hemiplegia.

5) Weber's syndrome: Ipsilateral third nerve and contralateral hemiplegia.

h) Vestibular + cochlear symptoms are indicative of cranial nerve 8 disease at the level of the labyrinth or of the cranial nerve 8. If other symptoms are present, brain stem may be involved.

7. Cerebral Cortex

a) Focal (partial) seizures are always indicative of disease involving the cerebral cortex. The specific clinical phenomena of the seizure will depend on the location of the lesion. Rapid secondary generalization of the seizure discharge may obscure the origin.

b) Lateralized deficits in terms of upper motor neuron findings and cortical sensory deficits (somatosensory or visual) may occur based on the location of the lesion.

c) Aphasia is always indicative of involvement of the dominant (usually left) hemisphere, in particular involvement of the speech areas or of the fiber systems inter-connecting the speech areas.

d) Cranial nerve involvement of a lower motor neuron type does not occur. Corticobulbar involvement may produce supranuclear weakness of the lower half of the face. Bilateral corticobulbar damage may produce a pseudobulbar state involving cranial nerves 5; 7, 9, 10, 11and 12.This is manifested by a hyperactive jaw jerk, spastic speech and emotional lability.

e) Changes in personality and behavior are indicative of disease involving the cerebral cortex particularly the frontal and temporal lobes.

f) Changes in immediate (working) memory often reflect neocortical pathology particularly involving the prefrontal areas. Changes in the ability to consolidate new learning may reflect limbic pathology (medial temporal and medial thalamus).

8. Basal Ganglia

a) There is no direct involvement of the lower motor neuron.

b) Major upper motor neuron descending motor pathways such as the pyramidal tract are not affected.

c) A modulating circuit is dysfunctional. This dysfunction is manifested by alterations in motor function characterized by the following findings:

1) A lack of movement, akinesia or slowness of movement, bradykinesia

2) Excessive movement: tremor or dyskinesia

3) Increased resistance to passive motor: rigidity

4) Alteration in righting reflexes affecting gait and balance

d) No sensory symptoms are present

e) No direct involvement of cranial nerve function is present but the motor effects described above may alter cranial nerve function

f) Cognitive function is usually not directly involved although, many diseases that affect the basal ganglia also affect cerebral cortex or frontal basal ganglia connections and thus are

associated with changes in mental status.

9. The Cerebellum

a) Lower motor neuron function is not affected.

b) Upper motor neuron function in terms of long tracts is not affected.

c) A modulating circuit is dysfunctional. The clinical effects of this dysfunction depend on the area of cerebellum involved.

1) Lateral cerebellar hemisphere relates to the ipsilateral arm and leg. Deficits produce dysmetria of the arms and legs: intention tremor, finger-to-nose and heel-to-shin deficits and an incoordination of movements.

2) Midline cerebellum relates to the axis of the body. Deficits produce ataxia of trunk.

3) Floccular nodular (archicerebellum) relates to the vestibular system. Deficits produce a loss of balance in sitting and standing

d) Sensory function of a conscious nature is intact.

e) Cranial nerve function is generally intact, although speech and eye movement are altered by cerebellar dysfunction. At times vestibular function may be altered

f) Certain aspects of cognitive function and motor learning are altered.

C. THE NATURE OF THE PATHOLOGY:

1. The Concept Of Extrinsic Versus Intrinsic: Extrinsic diseases are usually focal mass lesions compressing the spinal cord or brain. Intrinsic disorders arise within the substance of the nervous system. Disorders of muscle, and neuromuscular junction are usually not due to compressive disorders.

2. The common pathological processes related to the site of lesion.

a. At the level of muscle, the common processes are (1) degenerative disorders on a genetic basis: the dystrophies primarily occurring in children (2) genetic congenital myopathies, (3) the metabolic myopathies, often with a genetic basis, (4) the acquired inflammatory myopathies related to auto immune disorders.

b. At the level of the neuromuscular junction, the most common disorder is myasthenia gravis, an autoimmune disorder in which an antibody to the acetylcholine receptor blocks and damages the postsynaptic receptor. Other disorders, involve a paraneoplastic related antibody to calcium channels, which alters the release of acetylcholine from the pre synaptic sites. Botulinum toxin produced by an anaerobic bacterium also alters transmission at the neuromuscular junction.

c. At the level of peripheral nerve, the most common causes of a mononeuropathy are direct trauma and the entrapment syndromes in which compression of a nerve occurs. Vascular disease may also produce a mononeuropathy. As regards polyneuropathies, the most common causes are diabetes mellitus, and B vitamin nutritional deficiencies, followed by toxic and hereditary disorders. In parts of the world the infectious disorder leprosy is the most common cause of a multiple mononeuropathy or of a poly neuropathy.

d. At the level of the nerve root, the most common disorder is compression due to ruptured disks or to the osteophytes of the degenerative disk disorder cervical spondylosis. Tumors (Schwannomas) may also arise from nerve roots and peripheral nerve and in the process compress the nerves.

e. At the level of the spinal cord extrinsic disorders are the more common disorders. The mass may be a ruptured disc, a tumor in the epidural or intradural space, collapsed vertebrae due to metastatic disease, trauma or infection, or an infectious process such as an epidural empyema. Intrinsic processes may be focal, system disorders or multifocal disorders. Examples of focal intrinsic processes are infarcts due to anterior spinal artery occlusion, intrinsic tumors such as gliomas, or a focal enlarging cyst as in syringomyelia or transverse myelitis. Examples of system disorders are amyotrophic lateral sclerosis due to the degeneration of the motor neuron in the anterior horn or posterior lateral column disease of vitamin B12 deficiency. An example of a multifocal disorder is multiple sclerosis, which is the most frequent of the

intrinsic disorders.

f. At the level of the brain stem, most disorders are intrinsic. The most frequent disorder is vascular disease. The major type is ischemic occlusive involving the vertebral-basilar arteries producing infarcts. Other intrinsic disorders include tumors, hemorrhage, and demyelinating disease. However the brain stem may be affected by extrinsic processes arising in cerebellum (tumors and hemorrhages) or by tumors arising from the Schwann cells in cranial nerves (primarily the vestibular nerve).

g. At the level of the cerebellum most disorders are intrinsic. Infarcts secondary to occlusive disease are the most common. Other disorders are due to hemorrhage, neoplasms, degenerative disease and multiple sclerosis. In infants and children, tumors are the most common disorder.

h. At the level of the diencephalon, most disorders are intrinsic with the exception of tumors arising from the pituitary and secondarily compressing the hypothalamus. The most common intrinsic disorder is vascular disease (infarcts and hemorrhage). Other disorders are gliomas, and nutritional (Wernicke's encephalopathy due to thiamine deficiency). In children neoplasms are the most common disorder.

i. At the level of the basal ganglia, all disorders to be considered are intrinsic with degenerations being the most common (Parkinson's and Huntington's disease). However, the basal ganglia are also the most frequent site of intracerebral hemorrhage related to the penetrating branches of the middle cerebral artery. These lenticulostriate branches are also subject to occlusion producing infarct involving the internal capsule and basal ganglia.

j. At the level of cerebral cortex and subcortical white matter, the common pathological processes are intrinsic and relate to the age of the patient. In infant's hydrocephalus or congenital malformations or migration disorders may affect cortical function. In children, adolescents and young adults, trauma is a major consideration. In the adult, intrinsic tumors of the glial series are a major consideration. In the older patient, ischemic-occlusive vascular disease is a major neuropathologic process. In the elderly patient, the degenerative disorder, Alzheimer's disease afflicts a significant proportion of the population. In all disorders involving the cerebral cortex, recurrent seizures (epilepsy) are major considerations at all ages. Extrinsic disorders include the previously mentioned head trauma and the extrinsic tumor, the meningioma.

Part II — Neurological History and Examination will be found on CD-ROM.

Part III — Diagnostic Studies in Neurology will be found on CD-ROM.

PART 2: THE NEUROLOGICAL HISTORY AND EXAMINATION

The neurological examination provides a means for the systematic analysis of symptoms and signs.

I. AN OUTLINE OF THE COMPLETE NEUROLOGICAL HISTORY & EXAMINATION

[Note - that for each patient the total history and examination may be tailored so that for some cases abbreviated evaluations may be carried out in a particular area.]

HISTORY: The importance of the detailed history cannot be overemphasized, 75% of neurological diagnosis is dependent on the history.

1. Demographic Data: Age, sex, marital status, handedness, occupation, level of education.

2. Chief Complaint.

3. Present Illness.

4. Review of Symptoms Relevant to the Neurological System:

a. Mental Status: Orientation, memory, personality changes, mood changes, delusions, hallucinations, (see below regarding witnesses)

b. Language function.

c. Loss of consciousness: Syncope, Seizures: Details as to the onset, course, duration, confusion or other neurological symptom before, during and after. If relevant may need to obtain information from witnesses and/or relatives, friends, etc.

d. Alterations in alertness and sleep patterns.

e. Cranial Nerves:

I - Anosmia

II - Blurring or loss of or distortion of vision

III, IV, VI - Double vision

V - Numbness or pain in the face

VII - Weakness of the face, alterations in taste

VIII - Decrease, loss of or distortion of hearing, Tinnitus, vertigo, dizziness + nausea & vomiting

IX, X: Dysarthria: (change in voice, hoarseness); dysphagia (Difficulty in swallowing).

XII - Alterations in tongue movements

f. Motor system: Weakness, incoordination, clumsiness, unsteadiness - in sitting, standing, walking

g. Alterations in sensation: paresthesias, dysesthesias, tingling, and numbness

h. Strokes

i. Headaches

j. Trauma to head, neck, back or extremities.

k. Bladder and bowel function: incontinence, frequency, and spasticity.

5. General Medical History

a. System Review

b. Hospital admissions

c. Surgical history

6. Family History

7. Social History

EXAMINATION:

General Physical Examination to include vital signs

Neurological Examination

1. Mental Status

a. Level of consciousness: alertness

b. Orientation: time, place and person

c. Information: presidents, capitals, historical or local or sports or political information.

d. Memory:

1. Immediate recall (working memory): digit span and repetition of words.

2. New learning - at times, referred to as short term or test of labile phase of remote memory: delayed recall 5 out of 5 in 5 minutes or 3 out of 3 in three minutes.

3. Remote recall: date of birth, marriage, names and ages of children, siblings, parents, etc.

e. Insight as regards illness, etc.

f. Abstract reasoning -

1. Similarities: concrete versus abstract

2. Proverb interpretation: concrete or abstract

g. Calculations: simple additions, subtractions, problems and serial 7s subtractions.

h. Language and Related Functions:

1. Fluency

2. Naming of objects

3. Repetitions

4. Ability to follow spoken and written commands

5. Reading

6. Writing

7. Arithmetic

8. Drawing: constructions

9. Apraxia testing

10. Left right orientation

11. Recognition of objects, pictures

i. Mood, affect, how appropriate, level of anxiety

j. Observed hallucinations and perceptual distortions

2. Observed seizure activity describe in detail

3. Cranial Nerves:

a. I: Sense of smell

b. II: Fundi, visual fields, blind spot, acuity

c. III, IV, VI:

1. Pupillary responses: light, accommodation direct and consensual

2. Extraocular movements

3. Nystagmus: spontaneous, induced by eye movement, caloric stimulation, Hallpike maneuvers

d. V:

1. Sensation: Touch and pain

2. Jaw movement

3. Jaw jerk

e. VII:

1. Facial movements: upper and lower face

2. Labial sounds - dysarthria

3. Taste

f. VIII: Auditory tested. Vestibular if indicated as in coma, test calorics.

1. Hearing: Whisper perception; watch tick

2. Weber, Rinne

g. IX, X:

1. Movement of palate

2. Sensation of pharynx

3. Gag reflex

4. Guttural sounds dysarthria

h. XI: Sternocleidomastoids, trapezii: strength against resistance

i. XII: Tongue

1. protrusion, lateral movements, fasciculations and fibrillations

2.Lingual sounds - dysarthria

4. Motor System:

a. Atrophy and fasciculations

b. Motor power: Grade 0-5: Note pattern of weakness: hemiparesis, paraparesis, distal, proximal

c. Tone: Flaccid, spastic, and rigid

d. Posture: sitting, standing with eyes open and closed (Romberg test)

e. Gait:

1. Standard

2. Heel to toe, tandem gait

3. Accessory movements

f. Coordination:

1. Finger to finger to nose

2. Heel to shin

3. Rapid alternating hand movement

4. Foot tapping

g. Spontaneous movements:

1. Fasciculations

2. Tremor - at rest; maintained posture, or on movement

3. Chorea, athetosis, myoclonus

4. Dystonia

5. Reflexes:

a. Deep tendon stretch: Grade 0-4 at biceps, triceps, brachioradialis, patella and Achilles.

b. Superficial

1. Abdominal

2. Plantar: Sign of Babinski and associated reflexes

c. Frontal release signs: Grasp, suck, palmomental

6. Sensation:

a. Primary: pain, touch vibration

b. Cortical modalities: position, double simultaneous stimulation, tactile localization, stereognosis, graphesthesia.

7. Meningeal Irritation Signs:

Kernig's and Brudzinski

8. Vascular:

a. Carotid and temporal artery pulses and auscultation

b. Subclavian pulses and auscultation

c. Radial pulses and lower extremities (peroneal, and posterior tibials)

d. Bruits over head, orbits, vessels

9. Cervical Spine:

a. Range of motion: flexion, extension, rotation, and lateral displacements.

b. Local tenderness

c. Supraclavicular tenderness

10. Thoracic Spine: Local tenderness

11. Lumbar Spine:

a. Local tenderness

b. Mobility for flexion and lateral motion

c. Sciatic and femoral tenderness

d. Straight-leg-raising and reverse straight-leg-raising

12. Peripheral Nerves: Palpation + tap

a. Occipital at occipital notch

b. Ulnar at olecranon groove

c. Median at carpal tunnel

d. Sciatic at sciatic notch

e. Femoral at femoral canal

f. Peroneal at fibular head

g. Post tibial behind medial malleolus

Note - tenderness, enlargement, Tinel's sign (tingling on palpation)

13. Examination of head, face, tongue, and mouth for bruises, lacerations, hematomas of the scalp.

14. Examination of the limbs and body for bruises and malformations: Cafe au lait spots, vascular nevi, etc.

Clinical Impression:

Anatomical location of lesion

Differential Diagnosis as regards pathology

Laboratory Data already available: if relevant

Conclusion and plan of diagnostic and therapeutic management

II: ABBREVIATED NEUROLOGICAL EXAMINATION

A. Mental Status and Language Function:

1. Alertness
2. Orientation
3. Delayed recall (5/5 objects in 5 minutes)
4. Naming of 5 objects
5. Repetitions: "No ifs, ands or buts"

B. Cranial Nerves II - XII: Fundi, pupils, EOM's, facial movement and sensation, tongue move and gag, shoulder shrug and and SCM on head rotation

C. Motor System:

1. Resistance against force at shoulder abductors, biceps, triceps, wrist extensors, hand grip and finger abductors; hip flexors, quadriceps, hamstrings, ankle and toe dorsiflexors
2. Walk a routine gait; walk a tandem gait; walk on toes and heels
3. Stand on narrow base, eyes open, eyes closed
4. Other cerebellar - finger to nose, alternating hand motions
5. Any atrophy or fasciculations
6. Any tremor or other extra movements noted

D. Reflexes:

1. Deep tendon reflexes at biceps, triceps, brachioradialis, patella and Achilles
2. Plantar responses

E. Sensation:

1. Pain and touch sensation - extremities, shoulders, body
2. Vibration and position at toes

III: Mini-Mental State Examination (MMSE)

(After Folstein, M.S., Folstein, S.E., and McHugh, P.R. "Mini-Mental State". A Practical Method for Grading the Cognitive State of Patients for the Clinician. J.Psychiatr.Res, 12:189-198, 1975)

points

1. What is today's date? Month___ Date___ Year___ Day of week?___ Season___ ___ (5)

2. Where are we? City___ County___ State___ Hospital___ Floor___ ___ (5)

3. Repeat after me: ball - flag - tree Record the number recited initially ___ (3) Repeat them up to six times for registration

4. Subtract 7 from 100: 93 - 86 - 79 - 72 - 65 Spell WORLD forward and backward: D - L - R - O - W (Write the greater of these two scores to the right) ___ (5)

5. Repeat the three words: ball - flag - tree ___ (3)

6. Read and obey ("close your eyes") ___ (1)

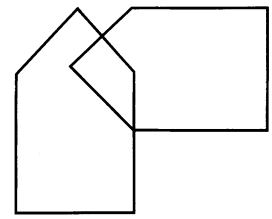
7. Name these items (pen and watch) ___ (2)

8. Repeat after me ("no ifs and or buts") ___ (1)

9. Take the piece of paper in your right hand, fold it in half, and put it on the floor. ___ (3)

10. Write a sentence: ___ (1)

11. Copy this design:



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PART 3: DIAGNOSTIC STUDIES IN NEUROLOGY

Specific laboratory studies are appropriate in providing information about disease affecting particular levels of the nervous system or about particular types of pathology.

A. Muscle and Nerve (fig 2-3):

1. EMG: Small needles are inserted into muscle to record the electrical activity of motor units. Under normal circumstances, at rest, no activity is recorded. With voluntary contraction, a significant number of units are recorded. These units have a range of amplitude and duration. In disease of muscle, voluntary contraction results in motor units of small amplitude and altered duration. In contrast when the anterior horn cell or peripheral nerve has been damaged resulting in denervation, spontaneous activity is present at rest. This consists of small amplitude fibrillations (the contraction of single muscle fibers) and **fasciculations** corresponding to the contraction of all the fibers in a motor unit innervated by a single anterior horn cell.

2. Nerve Conduction Velocity: Stimulation of nerves at two points along the nerve will allow the calculation of speed of conduction over specific segments. A specific site of block may be determined with normal conduction above and delayed conduction below that site, e.g. median nerve at carpal tunnel or ulnar nerve at olecranon groove of the elbow. Alternatively a general modification in conduction may be found in generalized peripheral neuropathies. Those peripheral neuropathies which involve primarily myelin produce a decrease in speed of conduction. Neuropathies that are predominantly axonal do not alter speed of conduction but may alter amplitude of the action potential. Repetitive nerve stimulation recording from appropriate muscle with measurement of the amplitude of muscle action potential may be utilized to study disorder of the neuromuscular junction. A progressive decrement occurs with myasthesia gravis and a progressive increment with Eaton-Lambert Syndrome.

B. Spinal Cord and Nerve Root:

1. Simple radiological studies: (fig 2-4)

Cervical spine, thoracic spine and lumbar sacral spine X-rays provide information about collapse of vertebrae, narrowing of spaces, narrowing of the neural foramina, metastatic involvement of vertebrae, fractures of vertebral elements and osteophyte (spur) formation. Neurofibromas may widen the neural foramina. Intrinsic tumors and syringomyelia may produce an increase in the diameter of the vertebral canal.

2. Computerized axial tomography scanning (CT scan) is now the most frequently employed technique in neurology but is less frequently utilized for spinal cord.

A computer is utilized to determine the differential attenuation of X-rays by the various tissues such as gray vs. white matter vs. blood vs CSF vs bone based on the differential content of water. An X-ray beam is passed through the tissues from multiple sites along a specific plane of section. The computer generates a series of slices usually in the horizontal plane.

Contrast enhancement is the technique of administering radio-opaque dyes intravenously of the types employed in intravenous pyelograms. These dyes do not usually cross the blood brain barrier. When this barrier is damaged as in tumors, brain abscesses, arteriovenous malformation, increased density will occur around or within the lesion. The barrier is also damaged in meningitis and around infarcts.

For the spinal cord; the most frequent use is in the lumbar area to image the nerve roots of the cauda equina.

3. Magnetic Resonance Imaging (MRI scanning) (fig. 2-5, 2-6) also utilizes computer-generated images. *MRI is now the study of choice for imaging spinal cord and nerve root.* Instead of utilizing X-rays, a strong magnetic field and radio frequency waves are employed. Placement of the patient's body in a magnetic field directionally orients the protons of that body. Passage of a brief radiofrequency current alters this directional orientation. When the radiofrequency current ceases the protons realign in the magnetic field. This realignment results in a signal. The signal as in CT scans, depends on the tissue density that is the differential water content of the specific tissue. Bone has little water content; gray matter and white matter have differential water content.

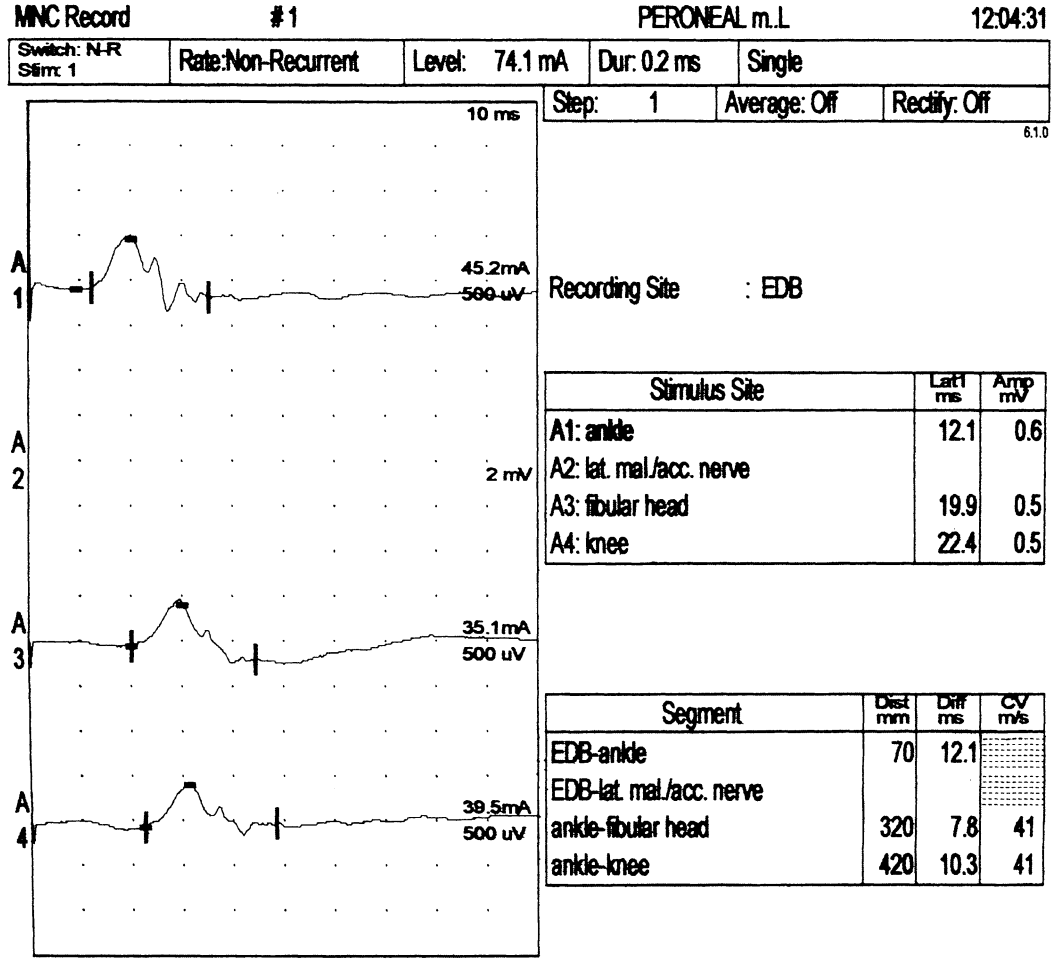


Figure 2-3 Motor nerve conduction velocity (normal). Various points along the course of the peroneal nerve were stimulated, with recordings from extensor digitorum brevis. The differences in latencies are calculated and then divided by the distance to yield a conduction velocity of 41 meters per second. (Courtesy Neurodiagnostic Laboratory University of Massachusetts Hospital)

Edematous and acutely necrotic tissue, as in infarcts and malignant tumors, usually has relatively high water content. In contrast to CT scan, images are obtained in the horizontal coronal and sagittal planes.

As in CT scans, contrast enhancement may be utilized. Gadolinium DTPA that normally does not cross the blood brain barrier is the agent employed.

The MRI procedure may be modified to vary the appearance of cerebrospinal fluid and of tissue water content. Altering the relaxation time (the interval after the application of the radiofrequency wave) will achieve this effect: Images obtained with a short relaxation time are labeled as T1 and emphasize the normal gray-white anatomical features with the CSF appearing as black. Images produced after longer relaxation times, labeled as T2, produce increased white appearance of CSF and other water content. Demyelinating lesions in multiple sclerosis are often prominent in T2 (MRI).

4. Other Radiographic Techniques Utilized in the Pre CT and MRI Era

Myelography: In this technique a radiocontrast dye is introduced into the subarachnoid space via a lumbar puncture. The spinal cord and nerve roots are then visualized by tilt-

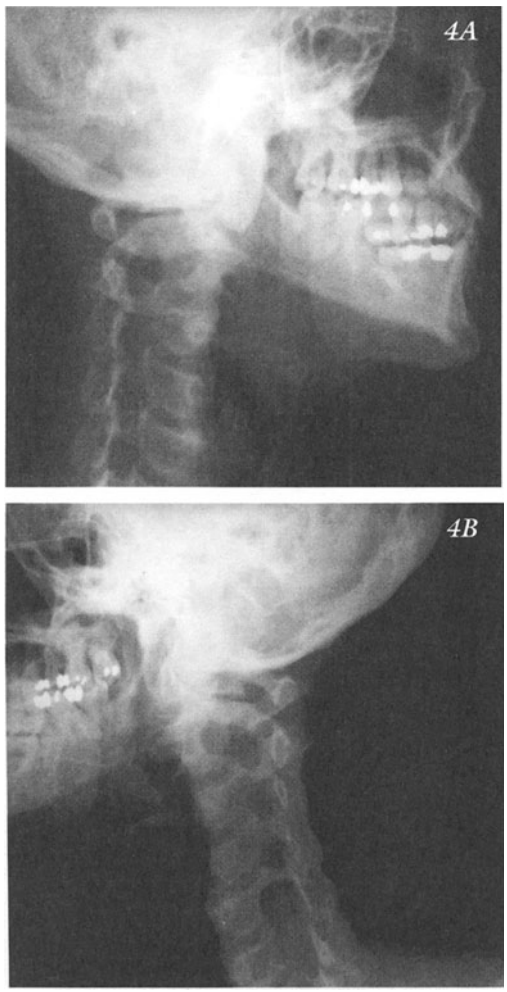


Figure 2-4. Cervical spine x rays. Marked Enlargement of the neural foramen has occurred on the left at C5-6 extending into C6-7 consistent with a neurofibroma/Schwannoma at this level. This 26-year-old female college maintenance worker developed numbness in the left arm extending from the elbow to all of the fingers and pain in the neck and left supra-clavicular area. She had absent deep tendon reflexes at left biceps and triceps. A) Right neural foramina on oblique view. B) Left neural foramina on oblique view. (Courtesy of Radiology Department Bay State Medical Center). See figure 2-5.

ing the body. Previously, oil soluble dyes (Pantopaque) were employed and had to be removed at the end of the procedure. Now water-soluble dyes are employed and these are absorbed. At times, CT scan is combined with myelography *(fig. 2-7)*. At times, the dye was allowed to enter the 4th ventricle or the cisterns around the brainstem.

5. Electrophysiological Techniques for Studying Spinal Cord Function:

a. *H. reflex*: submaximal stimulation of a mixed sensory-motor nerve at a voltage intensity which is insufficient to produce a direct orthodromic motor response (the M wave) will produce a muscle contraction after a long latency. This long latency response - the H. wave, involves the activation of the afferent fibers involved in the monosynaptic stretch reflex with activation of the anterior horn cell, anterior root motor fibers to the muscle.

b. *The F response*: Supramaximal stimulation of a motor sensory nerve produces an even longer latency response in the muscles. This depends on antidromic activation of motor neurons, which then induces, the longer latency discharge, activating the muscle fibers.

c. *Evoked Potentials:* There are computer-averaged signals associated with peripheral and central conduction following stimulation of specific sensory systems. The specific technique relevant to spinal cord is the somatosensory evoked potential. *(fig.2-8)*. Stimulation of median nerve in the upper extremity or of the posterior tibial or peroneal nerves in the lower extremities produces a series of waves. These waves are related to specific points in the conduction pathway. In the case of median nerve stimulation the waves relate to brachial plexus, cervical spinal cord and thalamocortical system. These studies are useful in detecting whether abnormalities are present in the posterior column/medial lemniscal system in multiple sclerosis or in spinal cord compression.

C. Brain Stem, Posterior Fossa and Skull Base

1. Radiological studies and other imaging studies:

a) *Skull X-rays* have in large part been replaced by CT scan and MRI scan but are still useful in providing information about skull fractures, enlargement of the pituitary, invagination of the odontoid and intracranial calcifications *(Fig.2- 9)*.

b) *CT scan* discussed previously may provide

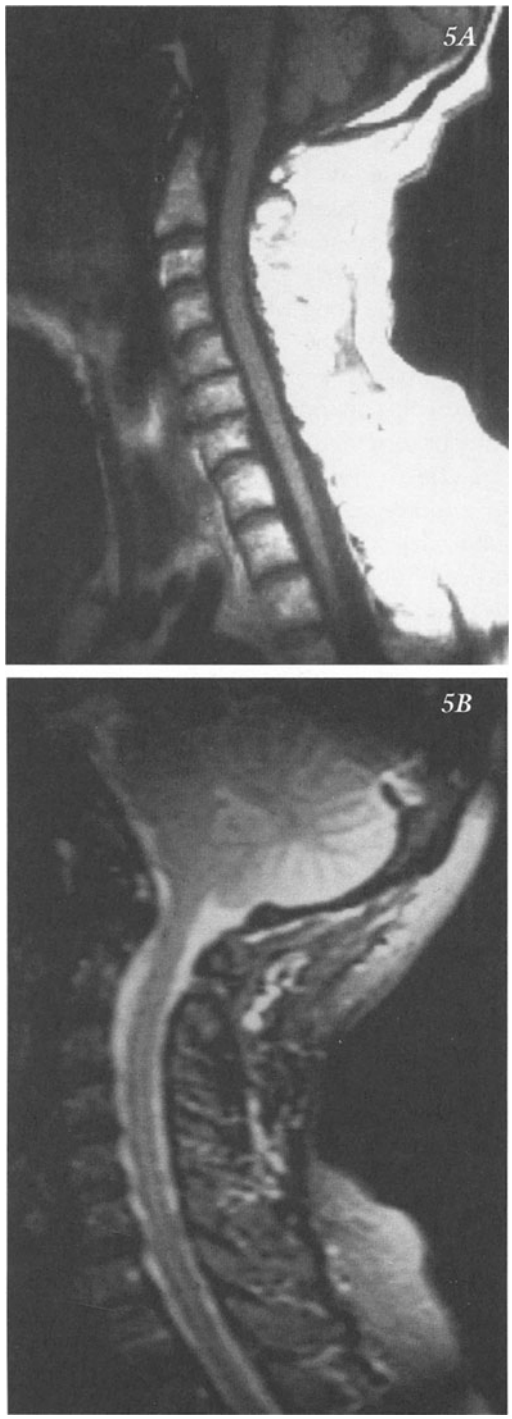


Figure 2-5. Magnetic Resonance Imaging (MRI) of the cervical and upper thoracic spine and spinal cord. A) T1 weighted B) T2 weighted from another patient: sagittal section close to midline. In both cases although degenerative disc disease is demonstrated, the spinal cord remains normal.

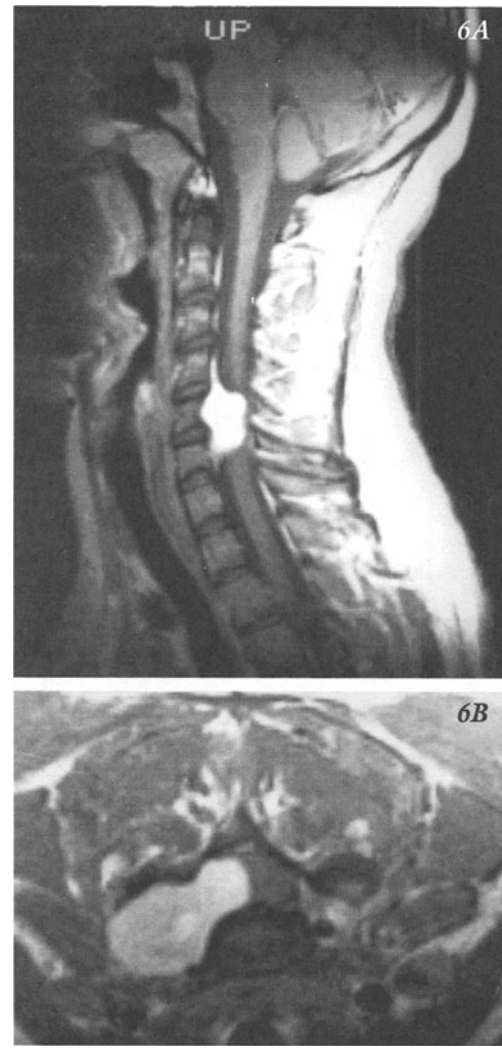


Figure 2-6 Cervical spine. MRI. Same case as figure 2-4.The cause of the enlargement of the neural foramina is now apparent .A large dumbbell tumor is present in the intra spinal bony canal and extends through the foramen to the extraspinal space. A) Sagittal view B) axial/transverse view. (Courtesy Radiology Department Bay State Medical Center)

information regarding tumors, infarcts and hemorrhages affecting the cerebellum and brain stem *(Fig 2-10)*.

c) *MRI* in general has become the standard technique for imaging the brain stem and cerebellum *(Fig 2-11)*.

2. Physiological Techniques

a) *Brain stem auditory evoked potentials* (fig. 2-12): A sequence of waves occurs which have been associated with specific points in the

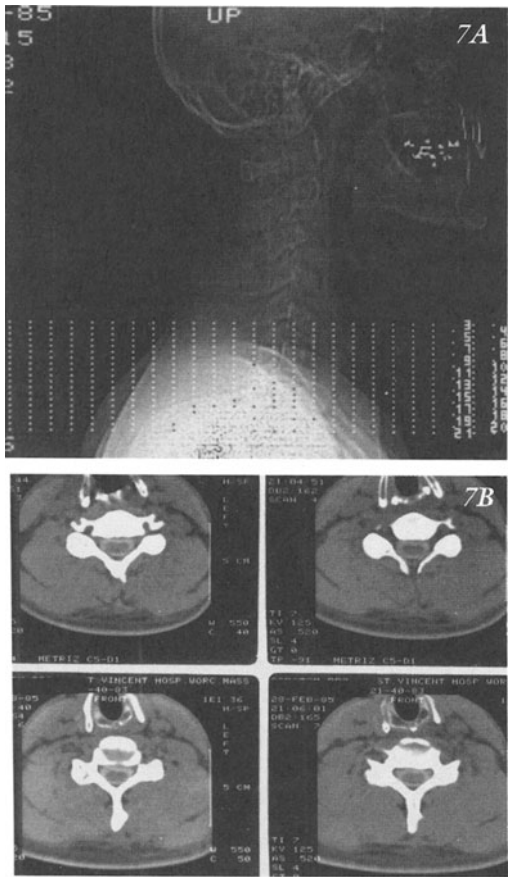


Figure 2-7 CT Computerized Axial Tomography (CT) after metrizamide myelography: normal spinal cord. A) Reference film for level of cross sections in this patient. B) Scans 3, 4, 6, 7- sections through C5-C6 Levels.

auditory conduction system:

Wave I; is associated with electrical activity generated in the cochlea at the origin of the auditory nerve.

Wave II; is associated with the entry of impulses from the auditory nerve into the cochlear nucleus at the medullary pontine junction.

Wave III; relates to signals generated at the level of the superior olivary nucleus in the lower pons.

Wave IV; relates to the nerve impulses generated in the lateral lemniscus.

Wave V; relates nerve impulses generated at the level of the inferior colliculus in the lower midbrain.

Delays are noted after wave I or between

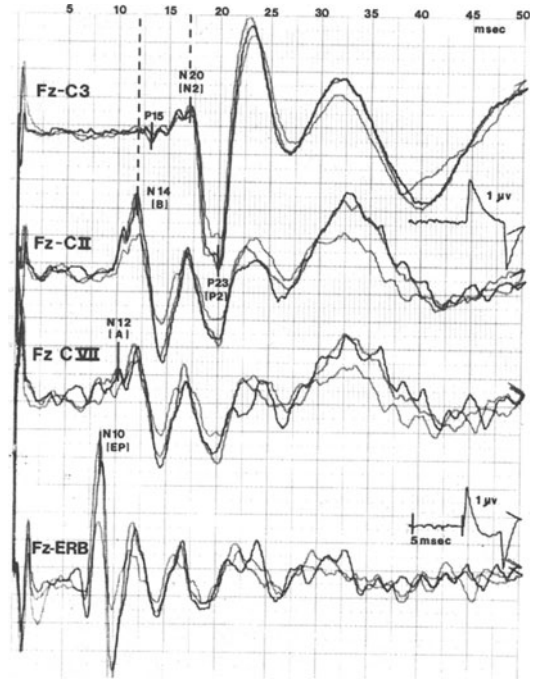


Figure 2-8: Short latency somatosensory evoked potentials (SER) median nerve stimulation at wrist. Recordings from supraclavicular area (FZ- ERB), cervical spine-VII (FZ-Cervical VII), cervical spine-II (FZ-Cervical II) and somatosensory cortical projection area and scalp (F2-C3). The specific wave forms-originate as follows: N10-Brachial plexus at Erb's point, N12-lower cervical spine-root entry area, N14-dorsal columns and dorsal column nuclei-lower medulla. N20-thalamocortical fibers and cortex. P23-Somatosensory cortex. (From Marcus, E.M. and Stone, B. In Evoked Potentials II, Ed. R.N. Nodar and C. Barber: Butterworth Publishers, Boston, 1984).

waves I and III in patients with acoustic neuromas (vestibular Schwannomas).

Somatosensory Evoked Potentials also provide possible information about conduction delays in the medial lemniscus.

b) *Special test of auditory and vestibular function:* audiograms, caloric testing an electronystagmograms.

3) Radiological Techniques No Longer Employed

a). *Pneumoencephalography* (PEG) *(Fig. 2-13):* Air was injected into the subarachnoid space via a lumber puncture. With the patient in the sitting position, the air would rise into the cisterns and ventricular system allowing

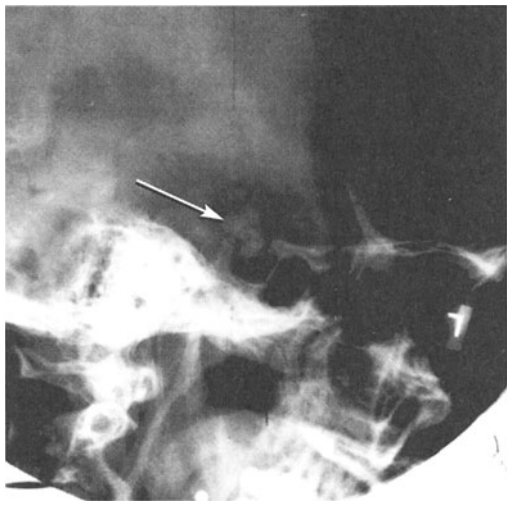


Figure 2-9: Skull X-ray Suprasellar Tumor: Craniopharyngioma. Enlarged sella turcica, and suprasellar calcification with changes in the anterior and posterior bony components (clinoids). This 23-year-old female had a six-year history of intermittent headaches and amenorrhea and recent diplopia. She had bilateral papilledema and an elevated serum prolactin level. Refer to figure 2- 13 (A) for a normal comparison (see also figure 27-14 for a CT scan of this case).

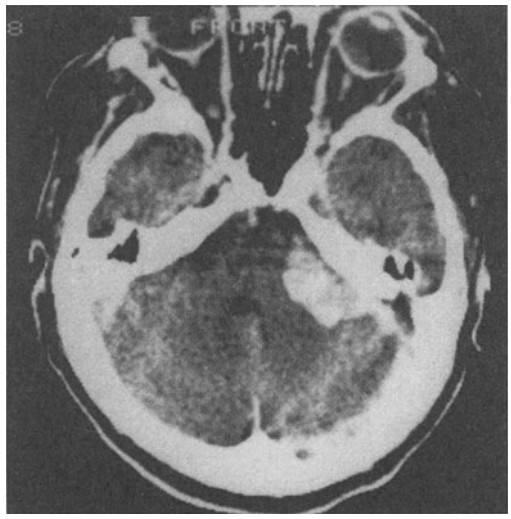


Figure 2-10 Computerized tomographic (CT) scan of the posterior fossa with contrast enhancement (C+). Cerebellar pontine angle tumor in a 65 year old female with a 17 year history of deafness in the left ear and a minor reduction of hearing in the right ear who had had additional findings of mild left peripheral facial weakness, ataxia of gait and dysmetria of left hand. The broad base along the left petrous bone suggested a meningioma. Courtesy of Dr. Tom Mullins.

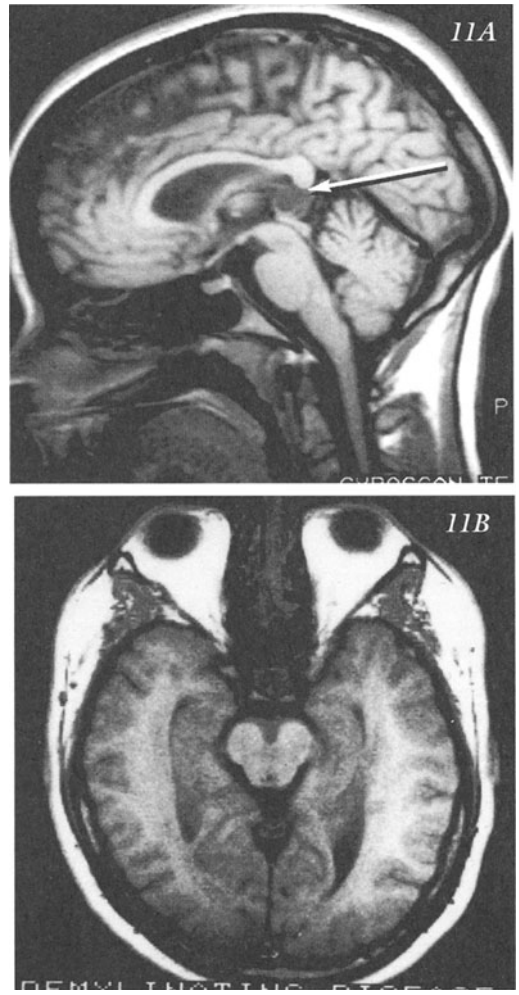


Figure 2-11. MRI: A) T1 sagittal section in a 29-year-old woman with multiple symptoms including headaches and depression but with no neurological findings. MRI/MRA was obtained because sister had an aneurysm and subarachnoid hemorrhage. This study reveals an apparently benign lesion of the pineal. B) MRI B) T1 horizontal /axial section in a young man with multiple sclerosis. No definite demyelinating lesions are demonstrated but the relationship of the mesial temporal areas to the mid brain is well demonstrated.

visualization of structures such as the fourth ventricle and aqueduct of Sylvius, as well as the lateral and third ventricle.

b. *Ventriculography:* In patients with increased intracranial pressure, or with tumors mass lesions in temporal lobe or cerebellum, the PEG was dangerous with the possible complication of herniation. Instead, a needle was introduced into the frontal horn and air or

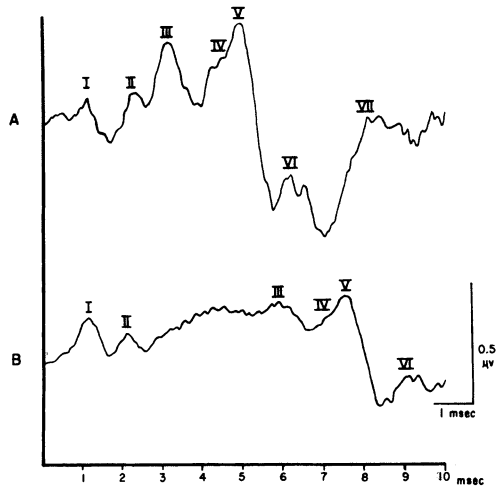


Figure 2-12 Brain Stem auditory evoked potentials (BAER). This 49 year old female had a progressive decrease in hearing in the left ear, decreased sensation on the left side of face for 2 years and additional findings of a left peripheral facial weakness and decrease pain sensation over the face. Imaging studies were normal but a delay was present in the II-III interval on the left suggesting a lesion between the cochlear nucleus and the superior olive. of unknown etiology. A) Normal right ear stimulation, B) abnormal left ear stimulation.

radio-opaque dye was introduced outlining the ventricular system and cisterns.

D. Cerebral Hemispheres

1. Imaging Techniques:

a) *CT scans* are usually employed in cases of acute trauma, intracerebral hemorrhage, subarachnoid hemorrhage, acute infarcts and acute brain abscess. In the case presented at the end of this chapter, an acute hemorrhage was demonstrated involving the leg and proximal arm areas. *(fig 2-29)*. (Refer to fig 1- for location of these areas on lateral surface of the cerebral hemisphere.

b) *MRI* - Has become the preferred technique for imaging patients with brain tumors, malformations, seizure disorders, inflammatory and demyelinating disorders *(fig. 2-14)*. In patients with ischemia and infarctions special diffusion weighted and perfusion studies are of value and are discussed in chapters 26.

Functional MRI - may allow correlation of metabolic activities and normal or disordered function *(fig 2-15)*.

c) As discussed above, pneumoencephalograms and ventriculograms are no longer employed.

d) *Radioisotope Techniques* have a limited value in selected cases.

1. *Radioactive Brain Scans* are no longer

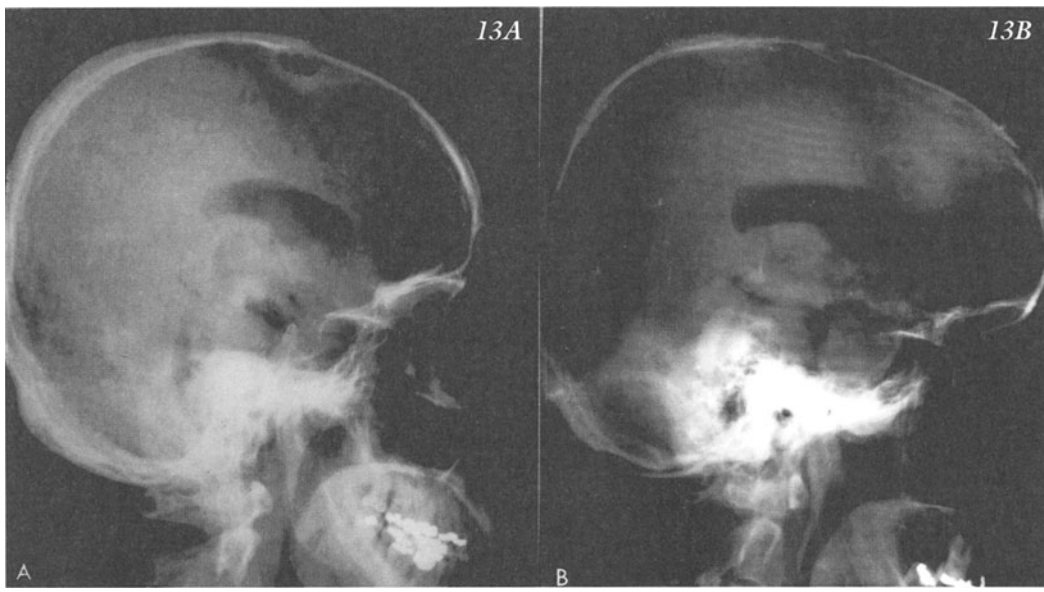


Figure 2-13. Pneumoencephalogram: A large pituitary adenoma has ballooned out the sella turcica on the X-ray. With the injection of air, via a lumbar puncture the extrasellar extension may be seen. A. Normal; B. Abnormal. (Courtesy of Dr. Samuel Wolpert, New England Center Hospitals)

performed. Prior to the development of CT and MRI this technique was of value in detecting lesions in which damage to the blood brain barrier or increased metabolic activity was present. Metastatic tumors, meningiomas, glioblastomas, abscess and acute infarcts were demonstrated but anatomical detail was poor. Several examples will be provided in the text

2. *Radioisotope flow studies* are still occasionally utilized to visualize the passage of a radioisotope injected into the lumbar subarachnoid space into the ventricles, out into the cisterns and through the subarachnoid space over the convexity to be absorbed into the venous sinuses. Normally, the isotope is no longer present in the ventricles at 48 and 72 hours. In the presence of communicating hydrocephalus (primarily normal pressure hydrocephalus), the isotope is still present in the cerebral ventricles at 48 and 72 hours (chapter 18 for an example).

3. *Single photon emission computed tomograph (SPECT)* scanning remains, as a modified form of the radionucleotide brain scan. Unlike the positron emission tomography scan a cyclotron is not necessary. Therefore cost is less, and availability is greater but resolution is poor *(fig 2-16)*.

4. *Positron emission tomography (PET) scan* employs positron emitting radio nucleotides combined with computer imaging of the emission to assess metabolic changes in specific areas of the brain. Resolution does yet reach the level of MRI or current high level CT scanning but now approaches that of early CT scans. Cost is high and availability limited since a cyclotron is required to produce the short life radioisotopes required. The major clinical use is in relation to focal (partial) epilepsy (Fig. 2-17). During seizure activity metabolic activity at the focus is increased. Between seizures the metabolic activity at the focus is decreased. The investigational use of PET scanning has provided valuable information about localization of function during the increased metabolic activity of normal cognitive activities, such as reading, motor activities, etc. In addition, information has been provided about the localization of altered metabolic activity in patients with schizophrenia, depression and various disorders of the basal ganglia. Functional MRI may provide a more effective technique for such metabolic correlations. Since positron emission tomography (PET scan) has not been previously considered in detail we will briefly review the technique at this point. This method combines CT with the use of positron emitting radioisotopes, which have been bound to compounds, which have significant biological function as metabolites or transmit-

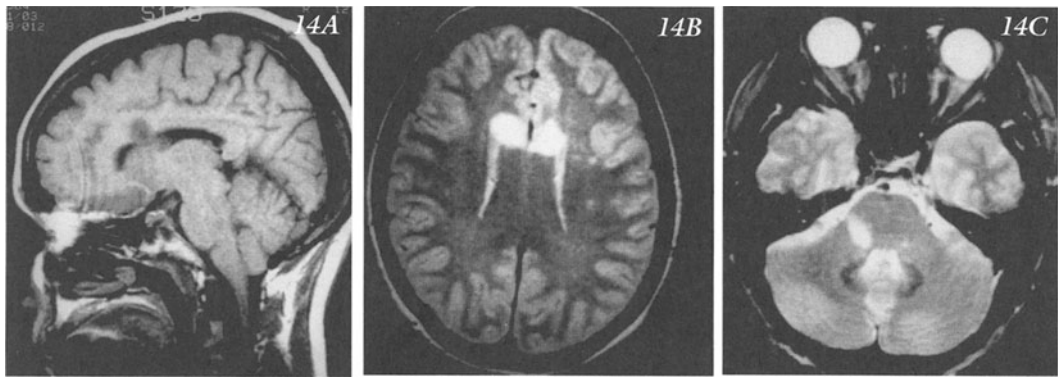


Figure 2-14. MRI. Multiple sclerosis. This 47-year-old female farm owner and manager had a 2-month episode of numbness ("novocaine type sensation") over the entire right trigeminal distribution. Five years previously she had intermittent unsteadiness Her examination demonstrated only a selective decrease in touch sensation over the entire right trigeminal distribution. The MRI studies confirmed a right mid pontine tegmental lesion but also indicated multiple areas of demyelination in the white matter of the cerebral hemispheres particularly involving the corpus callosum. A) T1 sagittal, (B) T2 horizontal views, (C) T2 axial of brain stem.

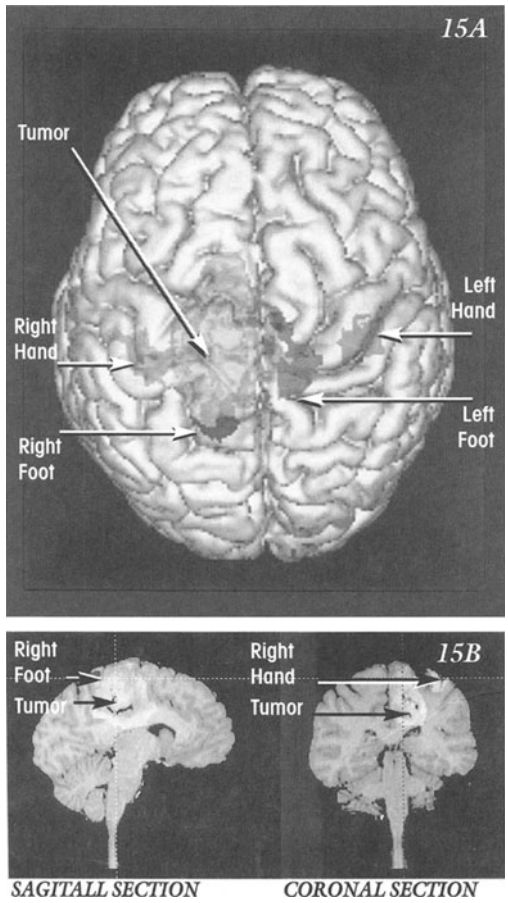


Figure 2-15. Functional MRI. This 33-year-old woman had a 2-year history of focal seizures involving the right foot and arm. A large area of the left premotor and motor cortex is involved by a grade 2 astrocytoma A) The relation of the areas activated by right hand or foot movements to the intrinsic brain tumor are indicated. B) Comparison of normal hemisphere to abnormal hemisphere. Note the displacement of the arm and leg areas by the tumor. Courtesy of Drs. B.R.Buchbinder, H. Jiang, G.R. Cosgrove A Cole, D. Hoch and R. Hill at the Massachusetts General Hospital Epilepsy Center.

ters. In contrast to standard CT scanning (where the source of radiation is the X-ray tube), in PET the positron-emitting isotope taken up by the tissue is the source of radiation. 2-Desoxyglucose is often employed since it is taken up by the neurons, and is phosphorylated as is glucose but is not further metabolized. The isotope of fluorine (18F) is bonded to the desoxy glucose. The uptake of glucose or of desoxy glucose into neurons is proportional to

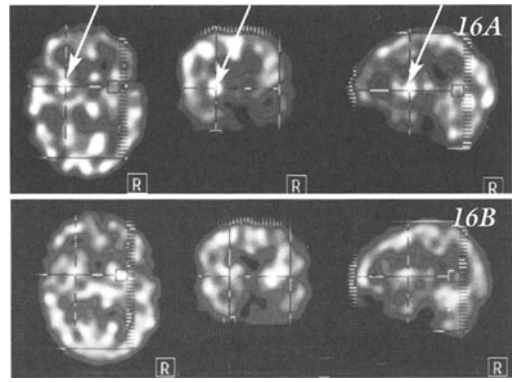
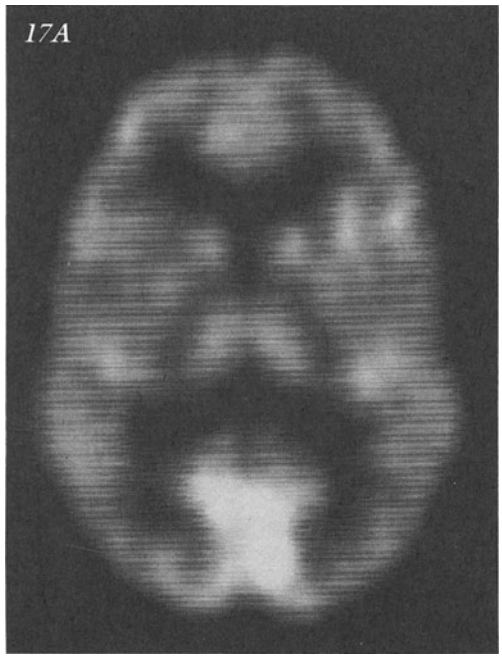


Figure 2-16. SPECT: Areas of increased perfusion during seizure activity are indicated in this scan. A) ictal uptake in areas of right temporal parietal and frontal lobes B) interictal. This 3-year-old child initially had infantile spasms with an EEG pattern consistent with the disorder hypsarrhythmia. Subsequently the EEG abnormalities were right anterior quadrant or parietal. Courtesy of Dr. Paul Marshall Pediatric Neurology University of Massachusetts.

the activity of the neurons. Thus, areas of cerebral cortex undergoing active seizure discharge will show increased activity and increased uptake. In normal individuals with activity of the visual system, e.g., opening the eyes to scan a scene, activity and uptake will increase in the visual projection area of the occipital lobe. With auditory stimulation, on the other hand, activity and uptake will increase in the auditory projection area of Heschl's transverse gyrus of the temporal lobe. Areas of damage will show decreased uptake.

2. Physiological Techniques: NOTE THAT SEVERAL ILLUSTRATIONS OF EEG RECORDS WILL BE FOUND IN AN EEG ATLAS SECTION OF THE CD ROM

a. *Electroencephalography.* This technique provides information about the electrical activity of the cerebral cortex. This activity represents primarily the summated activity of postsynaptic potentials generated in the cerebral cortex. The electroencephalogram in the normal awake adult resting with eyes closed is characterized by the *alpha rhythm (Fig. 2-18).* This rhythm is composed of a sequence of sinusoidal waves of 8-13 Hz (cps), which is maximal over the parietal-occipital recording



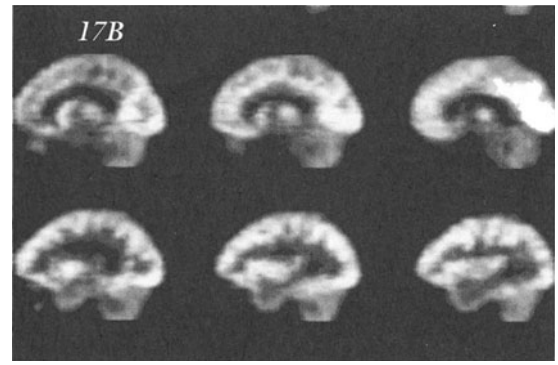


Figure 2-17. PET scans. The most prominent feature is the increased activity in visual cortex most likely reflecting visual activity during the study. MRI demonstrated mesial temporal sclerosis on the right and this study did demonstrate a possible decrease in activity right temporal area. This 22-year-old female had complex partial seizures since puberty. She had prolonged status epilepticus as an infant related to fever. A) Horizontal, B) sagittal. Courtesy of Dr. Cathy Phillips Neurology University of Massachusetts

area. Activity faster than alpha rhythm is referred to as *beta activity* and may be present over frontal areas. Increase amounts of beta activity occur as an effect of various drugs such as barbiturates and benzodiazepines. Alterations occur in this normal background activity related to the following factors: (1) eye opening producing reduction of amplitude *(Fig2-18C)*: (2) *age*: slower activity with infancy and childhood (*delta* 0.5- 3 Hz, *theta* 4-7 Hz), (Fig.2-19-CD ATLAS). (3) *Sleep, (Fig.2-*20- CD ATLAS), (4) *drugs and anesthesia:* faster and then slower activity (Fig.2-21-CD ATLAS)

Abnormalities may be (1) focal or (2) generalized.

1. **Focal abnormalities** may be categorized as:

A. *Focal spikes* implying focal excessive neuronal discharge involving the cerebral cortex and associated with partial (focal) epilepsy. (See chapter 29).

b. *Focal slow wave* activity, which implies focal cortical damage, as in infarcts, brain tumors and brain abscess. *(Fig. 2-22).*

c. *Focal suppression* of activity implies non-active electrical tissue under the electrodes - this may be seen with a fluid collection (subdural or intra cerebral hemorrhage) or with total destruction of tissue. *(Fig 2-23.)*

2.**Generalized abnormalities:**

a. *Generalized discharges of spike or polyspike – slow wave complexes* are associated with various types of generalized epilepsy. These will be discussed in chapter 29.

1) *Generalized bursts of 3/second spike wave* complexes are associated with absence seizures previously labeled petit mal epilepsy.

2) *Generalized bursts of polyspike and slow wave complexes* are associated with myoclonic seizures.

3) *Generalized polyspike discharges* may be found in the tonic phase of the generalized tonic clonic seizures.

b. *Generalized slow waves* are associated with diffuse disorders: infectious, ischemic, toxic or metabolic encephalopathies *(Fig. 2-24, 25).*

c. *Generalized periods of suppression* imply a more serious type of diffuse dysfunction, as in anoxia or a deep stage of anesthesia. Figure 17-16 provides an example of a burst suppression pattern.

d. *Total suppression of activity* may be found when neocortical death has occurred - as in

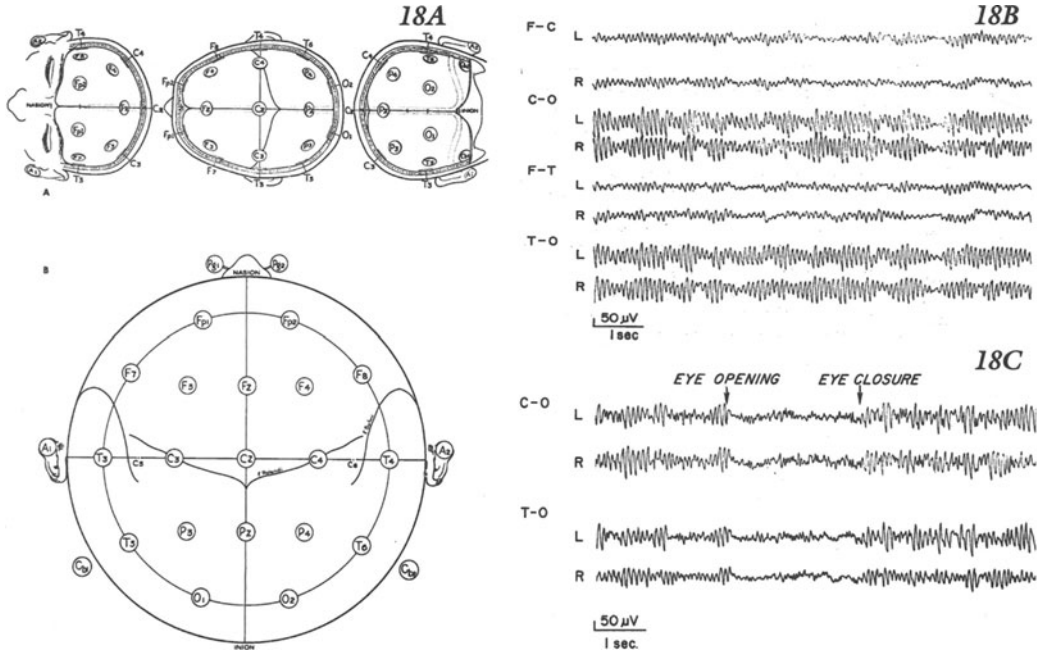


Figure 2-18. A) The International Federation Ten-Twenty Electrode Placement System. Frontal superior and posterior views; and a single plane projection of the head demonstrating the standard positions and the rolandic and sylvian fissures. The term ten-twenty is based on the placement of electrodes at particular percentages of the distance between nasion and inion. From Jasper, H.H.: Electroenceph. Clin. NeuroPhysiol. 10: 374, 1958 (Elsevier).
B) The normal adult electroencephalogram. The patient is awake but in a resting state, recumbent with eyes closed. F = frontal, T = temporal, C = central and 0 = occipital. (Bipolar recordings) These abbreviations will be utilized in subsequent illustrations. C) Effects of Eye Opening and Closure. Opening is associated with a blocking or suppression of the alpha rhythm of 10 cps and with the appearance of a low voltage fast-activity of 20 cps (beta rhythm). With eye closure, there is a return of the alpha rhythm.

severe anoxic encephalopathy or the vegetative state. This total suppression of activity accompanied by an absence of brain stem reflex activity and an absence of spontaneous respiration occurs in brain death. Similar findings of total suppression of activity both electrical and reflex may also occur under condition of deep anesthesia.

b. Specialized Techniques Employing Electroencephalography:

1. *Sphenoidal electrodes* - these are small needle electrodes placed in the sphenoidal sinus area to record from the medial aspects of the temporal lobe.

2. *Video EEG monitoring:* The recording of EEG and behavior onto single videotape allows correlation of electrical activity and of clinical seizure activity. This is usually a prerequisite study prior to epilepsy surgery and is also uti-

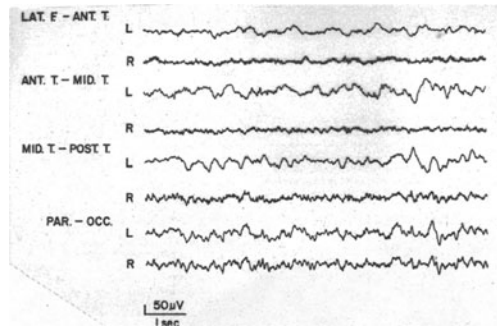


Figure 2-22. Focal 2-3 Hz slow wave activity in the electroencephalogram: left temporal area indicating focal damage in this area. Brain abscess, left temporal lobe in a 14-year-old female secondary to an acute S aureus mastoiditis extending into the petrous ridge. Electroencephalogram. Bipolar recording. (LAT. F. - lateral frontal; ANT- T- = anterior temporal; MID. T. = mid- temporal; POST. T- = posterior temporal; PAR. = parietal; OCC. = occipital). (Listed in text and CD ROM as Figure 27-22).

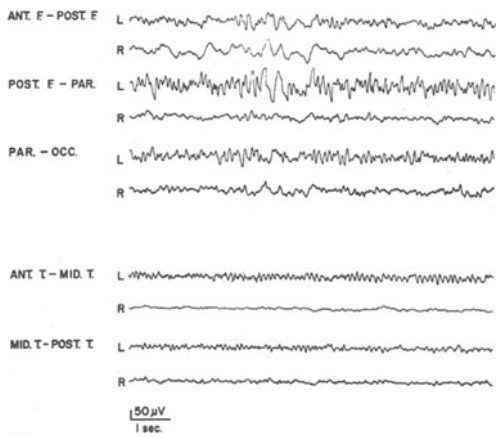


Figure 2-23. Focal suppression of EEG activity right temporal area and focal slow wave activity right frontal. Total right middle cerebral artery occlusion in a 61-year-old female with hypertension. Case 26-4. ANT.F. = Anterior frontal parasagittal; POST.F. = Posterior frontal (parasagittal); PAR. = parietal; OCC. = occipital; ANT.T. = anterior temporal; MID.T. = mid temporal; POST.T. = Posterior temporal. (Listed in text and CD ROM as Figure 26-14).

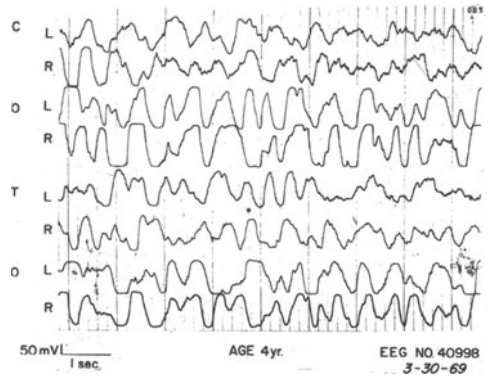


Figure 2-24 Generalized delta 1-2 Hz slow wave activity that persisted despite attempts at arousal this 4-year-old male had acute viral encephalitis.

lized in the analysis of pseudoseizures or other unresolved "spells".

c. *Depth electrode recording:* from medial temporal and other structures may be performed prior to epilepsy surgery often in combination with video monitoring.

d. *Subdural surface grids of electrodes:* may be placed on the cortical surface for better correlation of seizure discharges arising in the frontal or other neocortical areas.

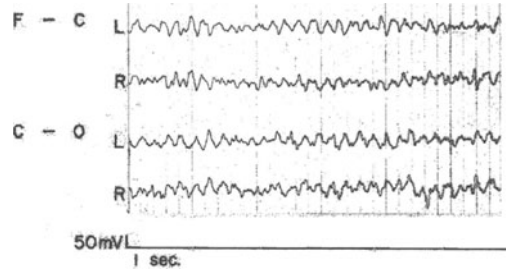


Figure 2-25. Generalized theta 5 Hz slow wave activity. This 67-year-old female had a metabolic encephalopathy due to impaired hepatic function secondary to cirrhosis and at this point was semicomatose in a stuporous state. When the patient was more deeply comatose, the awake activity was even slower at 4 Hz. When the patient was alert during intervening periods of recovery, the dominant activity was in alpha range.

e. *Electrocorticography:* recording directly from the pial surface may be utilized during epilepsy surgery.

f. *Polysomnography* (PSG): this technique is utilized in the evaluation of sleep disorders such as narcolepsy and sleep apnea. EEG activity from the parietal occipital or vertex areas of the scalp is correlated with

(1) Cardiac activity (EKG) rate and rhythm

(2) Respiratory activity rate and rhythm

(3) Oxygen (O2) saturation

(4) Extraocular movements

(5) EMG activity chins and or limb

The use of the PSG and the multiple sleep latency study will be discussed in chapter 29

c. Evoked Potentials:

(1) *Visual (VER or pattern reversal visual evoked potential: PVER):* The time of conduction over the entire visual pathway - to cerebral cortex is measured. Pattern reversal generates a prominent very stable wave at approximately 100ms, the P100 wave *(Fig.2-26).*

(2) Somatosensory evoked potentials: As discussed above, these studies may provide information regarding delays in conduction in the thalamocortical system.

3. *Neuropsychological tests:* A variety of tests have been developed. These include the Wechsler Adult Intelligent Score (WAIS) for-

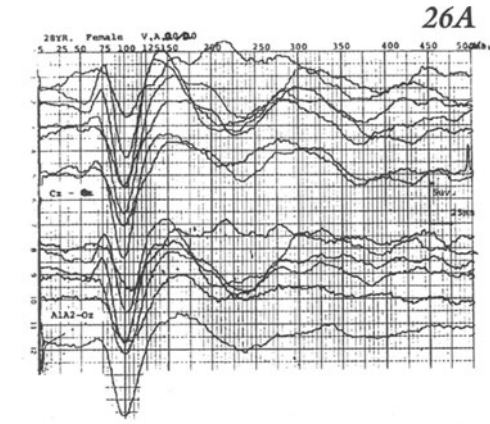
merly termed the Wechsler Bellevue Test of Adult Intelligence. This has a series of separate subtests covering multiple areas of verbal and performance functions. A total, verbal and performance intelligence quotients are derived. A series of tests have been developed to study aphasia and frontal lobe function and are discussed in those chapters. The Wisconsin Card Sorting Test is utilized to study cognitive perseveration. The Wechsler Memory Score provides a quantitative measure of memory function. The Minnesota Multiphasic Personality index, provides information regarding personality, affect, depression etc. Projection tests have also been developed to study personality function the Rorshark and the Thematic Apperception Test. The answers to the pictures provided unless very bizarre may be difficult to score and the results are open to several interpretations.

3. Techniques for the study of the cerebral circulation

a. Magnetic Resonance Angiography (MRA) - Normally in MRI scans, rapidly moving blood is not clearly imaged. However with special software programs, a non-invasive visualization of flow through vessels can be achieved. *(Fig 2-27, 2-28).* At present, resolution in the range of 2-3 mm can be achieved, allowing visualization of significant aneurysms. This procedure allows imaging of the carotid and other arteries prior to carotid endarterectomy*.

b. *In contrast, cerebral angiography (or arteriography)* is invasive. A catheter must be placed in the femoral artery and advanced into the aorta and then into the carotid or vertebral arteries. Radiopaque dye is then injected to directly image the cerebral vessels. This is the best technique when detailed study of the cerebral vascular is required, e.g., prior to aneurysm surgery. **With the increase resolution of MRA and of CT scan angiography, direct arteriography may be replaced by these non-invasive techniques.**

Under special circumstances, spinal angiography employing selective catheterization of



26B

PATTERN SHIFT VISUAL EVOKED RESPONSES

ANALYSIS P2 (P100) AT CHECK SIZE 4, 16, 64, 256, 1024, 4096 (CIRCLE)

LATENCY (MSEC) AS FUNCTION OF CHECK SIZE TOTAL CHECKS PER MONITOR SCREEN (VISUAL ANGLE OF ARC FOR EACH CHECK AT 1 METER)

SVH Controls N=42 Eyes 14-61 Years (Mean=30.8)	4 (7°)	16 (5°)	64 (2.5°)	256 (85')	1024 (30')	4096 (15')
Mean ± 1 SD	109.7±9	105.4±8	101.9±6	100.6±6	99.2±4.5	103.6±5
Range	93-130	93-120	92-117	90-114	91-109	95-117
Mean + 3 SD (P = .003)	137.3	129.1	120.7	117.7	112.7	118.7
Inter Eye Diff						
Mean ± 1 SD	4.4±4.1	5.4±5.8	2.4±2.1	2.1±1.9	2.7±1.9	3.2±2.8
Range	0-13	0-21	0-7	0-5	0-6	0-7
Mean + 3 SD (P = .003)	16.6	22.9	8.5	7.9	8.4	11.6

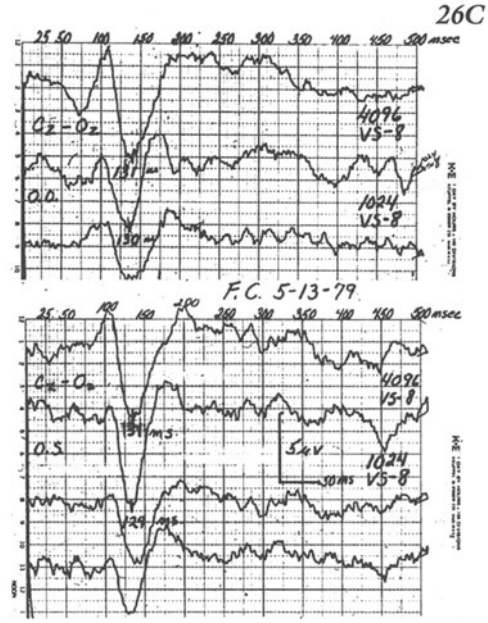


Figure 2-26. Visual evoked potentials. A) Normal. Each tracing represents the summation of 256 trials at different check sizes. Note the stability of the P100 response. B) Normal and abnormal values are obtained by statistical analysis with abnormal defined as >mean + 3 standard deviations. C) A patient with multiple sclerosis who had experienced several episodes of optic neuritis involving first one eye and then the other eye. Note the bilateral prolongation of the P100 responses.

radicular arteries may be performed to visualize spinal cord arteriovenous malformations.

c. *Duplex scans of the extracranial carotid and vertebral arteries:* this technique combines Doppler and ultrasound techniques to image blood flow in the major extra-cranial arteries.

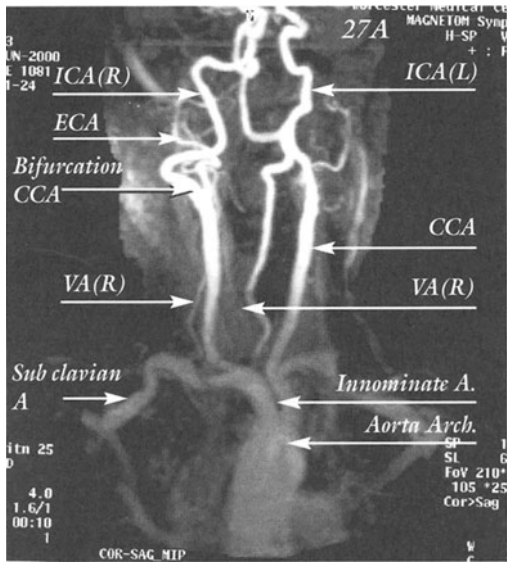
d. *Transcranial Doppler* - may provide gross information regarding flow in the major intracranial vessels.

CEREBROSPINAL FLUID (CSF) EXAMINATION

CSF fluid is usually obtained by a lumbar puncture. The lower end of the spinal cord, the conus medullaris, does not extend below the L2 vertebra. Therefore, introduction of a needle into the subarachnoid space between the L2-L3, or L3-L4, or L4-L5 spinous processes will not damage the spinal cord. CSF pressure when the patient is relaxed, but positioned on one side, will be usually less than 150 mm of CSF. Values in the relaxed state greater than 200 mm are considered abnormal. Respiration, abdominal pressure, flexion of head on chest or thighs and knees onto abdomen will all increase the pressure.

Normally no significant red blood cells (rbc's) should be present. When the puncture is traumatic the first tube collected will contain red cells but these should significantly decrease by the time that the fourth tube is collected.

Normally, less than 7 white blood cells (wbc's) should be present and all should be mononuclears. Any polymorphonuclears are abnormal and should raise the question of infection or inflammatory reaction. Spinal fluid glucose should be no less than 50% of the



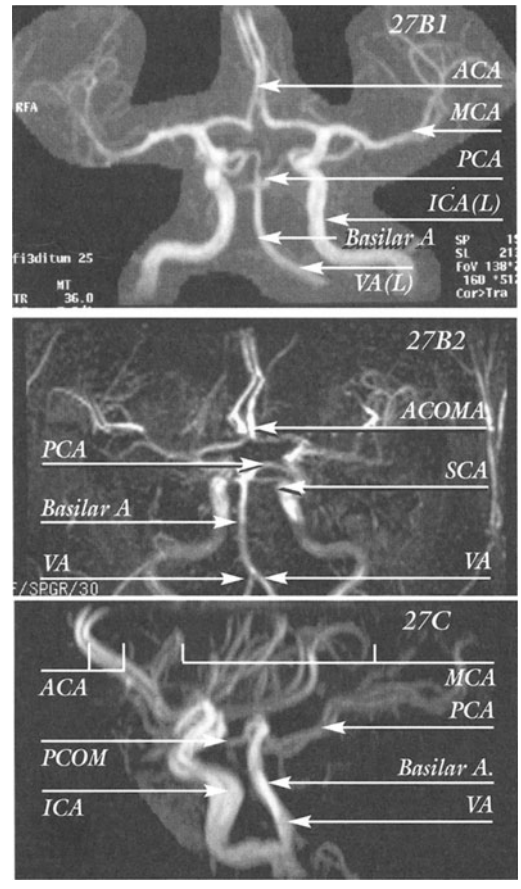


Figure 2-27. Magnetic resonance angiography (MRA). A) Aortic arch and major arteries in the neck. B1 & 2) Intracranial circulation coronal submental view demonstrated in two patients 27-B2 labels only the differences from 27-B1 C) Intracranial circulation (lateral view).
ACA= Anterior Cerebral A.
ACOM= Anterior Communications A.
CCA= Commono carotid.
ECA= External carotid A.
ICA= Internal carotid A.
MCA= Middle cerebral artery.
PCA= Posterior cerebral artery.
PCOM= Posterior communications
SCA= Superior cerebral artery.
VA= Vertebral artey.

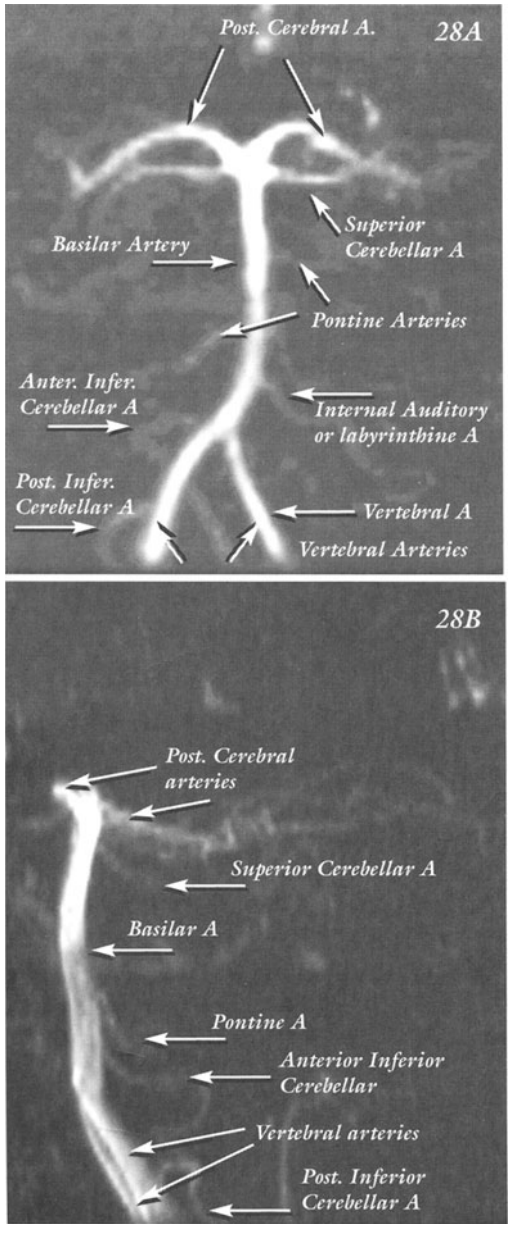


Figure 2-28.Magnetic resonance angiography. Basilar vertebral (posterior) circulation. A) AP view. B) Lateral view. The vertebral, basilar, posterior cerebral, and all of the circumferential cerebellar branches are evident.

blood glucose obtained at the time of the puncture or within two hours prior to the lumbar puncture. In acute bacterial meningitis - the CSF glucose is low due to the interference with the transport system and/or the increased metabolic activities.

Total CSF protein is usually less than 45 mg%. It is increased in a nonspecific manner in many processes affecting the nervous system.

a. Acute inflammation (meningitis and encephalitis).

b. Acute necrosis: infarcts, abscess and tumors

c. Blocks in the CSF - peripheral nerve barrier as in Guillain Barré syndrome. In the latter case no cells are present. This is referred to as albumin- cytologic dissociation and may also be present in patients with diabetes mellitus or myxedema.

d. Blockage of the CSF pathway at a spinal cord level. In patients with blocks in the lower thoracic or lumbar area, the protein level may be very high, with several grams present .The thick yellow fluid may clot in the test tube (Froin's syndrome)

The gamma globulin (IgG) percentage of the total protein may be increased under the following circumstances

1. Production of IgG in the serum is increased.

2. IgG is selectively produced within CNS by plasma cells - as in multiple sclerosis or neurosyphilis. Oligoclonal bands will also be present within the gamma globulin band.

ILLUSTRATIVE CASE HISTORY.

This patient should be compared to the patient of chapter 1. Both began with symptoms of weakness in the leg. However, the time course for evolution of symptoms differed significantly resulting in different diagnoses.

Case 2-1 This 90 year old right handed white male with a past history of hypertension awoke on the morning of admission with weakness of the left side, which was most marked in the leg. The initial admission examination indicated no motor function of the left leg, severe weakness of the left arm and minimal weakness of the left side of the face.

Past history indicated a previous minor "stroke" involving the left side of the body, from which he had made a full functional recovery with only minimal left sided weakness. During year prior to admission, occasional

periods of confusion had been present and progressive problems in memory had developed.

Neurological Examination: *Mental Status:* The patient was disoriented for time and place. He was however cooperative and able to follow all commands. He was fluent with no disturbance of language function. His remote memory was excellent. He could repeat the name of the examiner and of his primary physician but could recall neither name after five minutes. *Cranial Nerves:* A minor left central facial weakness was present. *Motor System:* He had no movement of the left leg. There was little function of the left shoulder and elbow. However handgrip was strong and independent finger movements were present the patient was recumbent in bed with external rotation of the left leg into a hemiplegic posture. *Reflexes:* Deep tendon stretch reflexes were absent in the lower extremities and the left upper extremity. The plantar responses were extensor bilaterally (bilateral sign of Babinski. *Sensory System:* No definite abnormalities within limits of testing.

Clinical diagnosis: 1. Cerebrovascular accident involving the superior parasagittal precentral gyrus either due to an anterior cerebral artery occlusion or a cerebral hemorrhage secondary to amyloid angiopathy. 2. Alzheimer's disease.

Laboratory data: A CT scan of the head demonstrated an acute hemorrhage in the superior parasagittal Rolandic area consistent with amyloid angiopathy. *(Fig. 2-29).*

Comment: This patient presents many of the neurological problems, which occur in the elderly. The patient had a one-year history of progressive memory problems, which primarily involved the formation of new memories. At age 90 such memory problems occur in more than 50% of the population. Usually, this represents the development of those degenerative changes in the neurons of the cerebral cortex seen in the process defined as Alzheimer's senile dementia.

The patient had elevated blood pressure for many years and had already experienced one "stroke" affecting the left side 10-15 years previously

In the present episode, the sudden development of symptoms would suggest an additional vascular event. Compare this case to Case 1-1 in which the patient had a gradual development of weakness in the leg secondary to a meningioma.

As regards the localization, the marked involvement of leg and proximal arm with relative sparing of hand and face might suggest a process involving the upper half of the motor cortex (refer to fig 18-12).

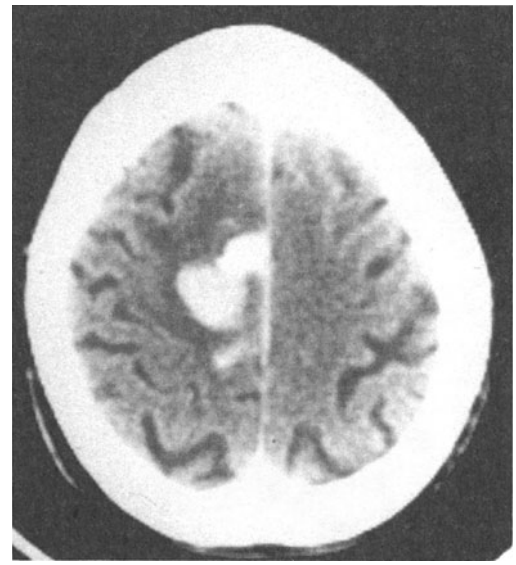


Figure 2-29 .CT scan. Cerebral hemorrhage in a 90-year-old man with left sided weakness primarily involving the right leg and shoulder. See case 2-1 at end of this chapter.