NEUROLOGY IN PRIMARY HEALTH CARE

Edited by

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Preface

The WHO Neuroscience Unit, Division of Mental Health and Prevention of Substance Abuse (MSA), has prepared this booklet for clinical neurological services at the primary health care (PHC) level.

The project is of great significance for the goal of the World Health Organization "Health For All By The Year 2000" which assumes that a reasonable quality of all aspects of health care, including neurological services, can be provided for all people by the target date.

Simplifying Neurology is however a difficult task. The strategy employed in the preparation of the booklet was to prepare a text directed mainly for services at primary health care level.

This booklet is now published for field trial.

We wish to acknowledge the technical and financial support of AIREN (Association Internationale pour la Recherche et l'Enseignement en Neurosciences), WHO Collaborating Center for Research and Training in Neurosciences, Geneva, Switzerland in the realization of this booklet.

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Chapter 1

NEUROLOGICAL HISTORY AND EXAMINATION

In Neurology, a complete history and a careful examination are essential for a correct diagnosis.

HISTORY

In taking the history, use a vocabulary easily understood by the patient, and listen to his complaints carefully. Note age, address, occupation and level of education.

Explore fully not only the neurological, but also the family, social and medical history of the patient:

- let him give his history in his own words; then, write down the more important points, and ensure that they are correct;
- enquire about personal habits: smoking, drinking, diet, drugs (medicinal and recreational);
- enquire about bladder, bowel and sexual functions;
- enquire about previous medical or surgical history: hospital admissions, surgical operations;
- ask about the family: age of parents or age and causes of death, if dead; age of siblings.
- the collaboration of a family member could be important especially with patients with impaired communication skills.

1) FAMILY AND SOCIAL HISTORY: what to ask for

1. Birth, development, scholastic history
2. Education, and vocation (for toxic exposure)
3. Family history of medical and neurological disease
4. Alcohol and drug use
5. Pregnancies and miscarriages
6. Smoking habits
2) **MEDICAL HISTORY**: especially ask for these diseases:

1. Hypertension  
2. Heart disease  
3. Stroke  
4. Head trauma  
5. Diabetes  
6. Endocrine disease (thyroid, adrenal)  
7. Cancer  
8. Medications  
9. Ophthalmologic diseases  
10. Other medical diseases

After having obtained a good history, review and enquire about the symptoms: onset, progression or regression, and duration. Let the patient describe them, and ensure that he understands the questions. After having obtained a full history, begin to review the neurological system: examine patient's vision, hearing, speech, reading, memory, intelligence and motor, reflex and sensory functions in the limbs.

**NEUROLOGICAL EXAMINATION**

The neurological examination is a complete and systematic examination of all functions of the nervous system. This allows the examiner to determine whether it is damaged and, if so, the nature and the location of the damage.

1) **MENTAL STATUS EXAMINATION**

During the examination, it is prudent to screen mental function rapidly: level of consciousness, behaviour, attention and concentration, memory, language, and abstract reasoning. (If abnormal, see Chapter 4).

![Mental Status Examination](image)
2) MOTOR EXAMINATION

a) Gait: observe the patient rising from a chair, walking in his normal fashion, and toe and heel walking. Difficulty may suggest weakness of the muscles of the legs due to brain damage, cerebellar disease, alcohol intoxication or peripheral neuropathy.

Watch arm and posturing: arm swing may be decreased on the side opposite to a damaged hemisphere.

In a spastic gait, the affected leg is thrown outwards from the hip and describes an arc. In ataxia, there is unsteadiness and a widened base in walking. In parkinsonism, the patient walks with few associated arm movements in a rigid fashion with a flexed trunk (Figs 1, 2 & 3).
b) Balance: one classical test for balance is the *Romberg test* (Fig. 4).

The patient stands with his feet together. And then he closes his eyes (Romberg sensibilized).

If the patient falls after closing his eyes, the test is considered positive: this usually indicates a damage to the spinal cord or a peripheral neuropathy, with loss of postural sensation.

A patient with a cerebellar lesion, on the contrary, is not able to control his position and movements, and sways excessively or falls whether his eyes are open or closed.

![Fig. 4](image)

![Fig. 4](image)

**c) Involuntary movements:**

*Athetosis and dystonia* are conditions characterized by slow, involuntary writhing movements of the trunk and extremities and generally associated with a fixed alteration in posture of the affected part of the body.

*Choreiform* movements are characterized by brief involuntary movements which may appear purposeful in character. There is often associated facial grimacing with movements of the tongue. These movements appear voluntary, but the person has no control over them. Chorea may be rheumatic and may result from previous infection with the streptococcus (Sydenham's chorea) or inherited, as in Huntington's disease.

*Tremor* is a rhythmic, repetitive involuntary movement, often of constant amplitude and frequency, generally involving one or more limbs, sometimes the head, neck, lips, tongue, or other parts of the body. It may be transient or constant, restricted or generalized, rapid or slow.

*Myoclonus* is characterized by a brief, shocklike contraction of a single muscle or of one or more muscle groups.

*Hemiballism* is usually of sudden onset, in elderly patients, giving rise to violent, writhing and choreic movements involving one side of the body and is usually related to damage to the subthalamic nucleus of the opposite side of the brain.
Tic is an abnormal movement which is intermittent, abrupt and aimless. It is caused by contraction of one or several muscles. The muscles of the face are commonly involved. Although tics may be psychogenic, they also occur often in the encephalitides and can occur in any type of brain pathology that involves the motor pathways in extrapyramidal disease.

Fasciculation is a form of involuntary contraction of groups of skeletal muscle fibers resulting in a localized twitching or flickering which can be seen under the skin or a mucous membrane but does not produce movement at a joint.

d) Limb tone (Fig. 5)
Muscle tone: passively flex and extend arms and legs, and elbow and knee joints. This allows one to evaluate muscle tone, that is the degree of resistance to passive movement.

Muscle tone may be increased (hypertonia), or decreased (hypotonia).

Hypertonic muscles are spastic or rigid; in both cases, there is an increased tone in antigravity muscles; in rigidity, the tone is increased in all the muscles of a joint, agonists and antagonists. Hypertonia may occur with damage to the brain due to vascular mechanisms, or with degenerative diseases.

Hypotonic muscles may offer less than normal resistance to stretch; they are flaccid. Hypotonia may occur with damage to motor nerve cells, cerebellum, peripheral nerves, or to muscles.

![The clasp-knife phenomenon of spasticity](image1)
![The cogwheel rigidity](image2)

**Fig. 5**
e) Strength: (Figs 6 & 7) muscle strength must be examined to elicit generalized or localized weakness.

Strength of arms and legs is evaluated by having the patient hold his arms extended out in front of him with his eyes closed; watch patient's limbs for some seconds: inability to maintain this posture on one side may be a sign of weakness.

In mild weakness (hemiparesis), the weak arm will pronate and drift downward. In more severe weakness, the affected limb will drift downward more rapidly.

![Fig. 6](image)

![Fig. 7](image)

Strength of muscles should always be tested on both sides of the body.

Muscle may be tested also by having the patient resist, and using adequate force to assess subtle weakness. Two tests with resistance may be used: patient's active motion against examiner's resistance, and patient's resistance against a movement performed by the examiner (Fig. 8).
As a general rule, symmetrical proximal weakness suggests myopathy (muscle disease), while distal weakness, upper neuron type of weakness suggests neuropathy (nerve disease).

Upper motor neuron type of weakness: an initial lesion affecting the upper motor neuron produces weakness on extension of the elbow and flexion on the knee.

f) Muscle bulk and consistency: they should be tested by palpating the muscles of the patients, and looking for tenderness, atrophy (wasting), or hypertrophy.

g) Coordination: this depends on an intact sensory and motor system.

In the finger-nose, heel-shin tests and testing for fine movements, observe the appearance of tremor (intention or action). Dysmetria is the inability to estimate the range of a movement, with past-pointing and overcorrection slowness of movement. Slowness of movement is seen with upper motor neuron damage and Parkinson's disease.

Incoordination ataxia may be related loss of postural sense (sensory ataxia), or cerebellar disease.
COORDINATION TESTS

*Finger-nose test*: the arm must be fully abducted, eventually to touch examiner’s finger, and then the patient must touch his nose with his finger. The movement must be repeated quickly. The test should be carried out with eyes open and shut (Fig. 9).

*Heel-knee test*: the patient must apply the heel of one leg to the knee of the other, and run it down the shin of the other, with eyes open and closed (Fig. 10).

Fine movements: have the patient touch his fingers successively to the thumb, first one hand and then the other. Compare the speed and accuracy of the movements. Impairments on one side are an indication of disease of the opposite side of the brain.

3) REFLEXES

a) *Muscle stretch reflexes*: they are tested when the examiner rapidly stretches a muscle, usually by striking the tendon with a reflex hammer, and observing the resultant muscle contraction (Figs. 11 and 12).
Reflexes may be increased, diminished, or absent; there is a great deal of variation in the activity of reflexes in normal individuals. For this reason, it is always important to compare the two sides.

Decrease or absence of tendon reflexes may result from any lesion affecting the reflex arc, such as peripheral neuropathies, or damage to the motor nerve cells in the spinal cord.

Increase of reflexes may result from a lesion of the upper motor neurons.

b) Cutaneous reflexes: Stroking the sole of the foot with a blunt object (eg. key); the normal response is flexion of the big toe. A pathological response is the extension of the great toe (Babinski sign) (Fig. 13).

Babinski sign is normally present in individuals under the age of about 12 months; after this age, it is an important indication of injury of the brain or of the spinal cord. It is usually associated with weakness, spasticity, and overactive tendon reflexes (muscle stretch reflexes).

4) SENSATION

It is best to test the patient's sensation with the patient keeping his eyes closed.

a) Touch: touch can be tested with a cotton swab; the patient must shut his eyes and say “yes” when he feels the touch.

b) Vibration: this is tested using a tuning fork over bony prominences, on each side of the body (Fig. 14). The patient must say whether he feels the vibration and whether it is of the same intensity on both sides, or proximally and distally in a limb.
Bony areas to be tested are: clavicle, elbow, wrist, dorsum of hands, fingers, anterior superior iliac spine, sternum, tibial tuberosity, lateral and medial malleoli, base of the big toe (Fig. 15). Begin with the fingers and toes. If vibration is felt there, it is not necessary to test more proximally.

Vibration sense tends to decrease in the toes and feet of elderly people.

c) Pin: stick lightly in each area tested; the patient must confirm whether he feels the pin-prick as painful or not. Then discard the pin.

d) Position: it is most easily tested using big toe and distal phalanx of the index or other fingers; the digit of one side is moved by the examiner through a very short excursion, up or down. The patient, eyes closed, must be able to detect even the slightest movement. Ask the patient when and in what direction the movement is perceived.

Total loss of pain sensation is called analgesia; impaired pain sensation, hypoalgesia; increased, hyperalgesia.
5) CRANIAL NERVES

- Smell (olfactory, 1st cranial nerve).
  Let the patient smell aromatic substances such as coffee, tobacco, and so on. Impaired olfaction (anosmia) may depend on several causes, not only neurological (smoking, rhinitis), and may be difficult to interpret. Do not test using irritant substances such as alcohol.

- Visual acuity (optic, 2nd cranial nerve).
  Acuity should be tested in each eye; blindness or loss of vision may be due to damage of the eye or of the optic nerve, or to injury of the brain; loss of visual acuity occurs in disorders of any part of the visual apparatus.

With one eye at the time, test ability to read letters or to distinguish figures of different sizes; usually test from a distance of 6 m.

**Visual fields**
Covering one eye, the patient has to fix on the examiner's pupil. The examiner moves his index fingers into the periphery of the patient's field until the patient himself reports first seeing them. The fingers (or the objects) should be brought in from all quadrants, that is at 1:30, 4:30, 7:30, and 10:30 on a clock face, to identify partial or complete field defects.

**Pupillary light reflexes**
This reflex is tested by shining a light directly into the pupil: the normal reaction is a pupillary constriction (miosis) (Fig. 18). It is an important test for assessing presence or absence of brainstem destruction in comatose patients. Each pupil must be tested separately.

![miosis](image)

Both pupils should constrict when a light is shone into either eye; if neither pupil constricts, the eye may be blind. If one fails to constricts, the oculomotor nerve (3rd cranial nerve) may be damaged or the eye impaired by previous injury or inflammation.

**Nerves tested by this reflex: optic and oculomotor.**
Look also for inequality of pupils: a dilated pupil on one side may mean nerve injury or brain swelling after injury; if this is the case, a more complete examination should be performed in an specialized hospital.
Eye movements (oculomotor, 3rd cranial nerve; trochlear, 4th cranial nerve; abducens, 6th cranial nerve).

To test ocular rotation, note any muscle imbalance when the patient looks straight ahead; then, have the subject look up and down, right and left, by following examiner's finger in the four directions.

Both eyes should move together; if one eye fails to show a full range of movements, a muscle or nerve may be paralyzed.

Diplopia or double vision is the symptom of the patient with paralysis of one or more of these nerves.

Corneal reflex (trigeminal, 5th cranial nerve; facial, 7th cranial nerve).

This reflex is tested by touching the cornea with a wisp of cotton wool over the iris. The response is a brisk bilateral eye closure, in defense.

Face (facial, 7th cranial nerve).

Look for asymmetry in the face, at rest and with movements: at rest, absence of wrinkles of one side of the forehead suggests paralysis of the facial muscles on that side; the corner of the mouth droops on the paralyzed side.

Three movements are important to demonstrate facial weakness:
- showing the teeth
- closing eyes tightly
- looking up (wrinkling the forehead) (Fig. 19).

Peripheral lesion of the 7th nerve causes weakness of all muscles of one side of the face; when the lesion is central, i.e. affecting the upper motor neuron pathway above the pons, there is a relative sparing of muscles of the eye and forehead.

Hearing (acoustic, 8th cranial nerve).

Look for auditory disturbances: a screening examination can be done at the bedside, testing each ear separately, using a tuning fork or simply rubbing fingers together near the ear.

Deafness may be due to middle ear disease, to disease of the cochlea, or the acoustic nerve, to otosclerosis, or to brain stem defects or damage. Vestibular tests are carried out if the patient complains of dizziness or unsteady gait.
Pharynx (glossopharyngeal, 9th cranial nerve; vagus, 10th cranial nerve).

Observe the pharynx and touch it on both sides with a wooden stick and observe the gag response (Fig. 20).

Fig. 20

A positive gag reflex is a contraction of pharynx, retraction of the tongue and elevation of the soft palate with elevation of the centrally situated uvula in the midline. This gag response may be absent in comatose patients, or sometimes if the nerves involved are injured.

Muscles of neck and shoulder (accessory, 11th cranial nerve)

The muscles involved are sternocleidomastoids and upper trapezius. The first is important in turning the head, and the second in shrugging the shoulders. Lesions of that nerve produce weakness in these muscles and impairment of the related movements (Figs. 21 and 22).

Fig. 21

Fig. 22
Tongue (hypoglossal, 12th cranial nerve).

Look for asymmetry, muscle twitching (fasciculation), or atrophy.

The tongue protrudes toward the side of weakness. Fasciculation indicates a lesion of the 12th nerve or of its nucleus.

6) MENINGES

Meningeal irritation may be revealed by testing the rigidity of the neck. Any pain or resistance when passively flexing the neck of the supine patient may be a sign of meningeal irritation.

In meningitis (meningeal infection or inflammation), the patient flexes his thighs when flexing the neck: this is called the Brudzinski sign (Fig. 23).

Fig. 23

Kernig sign: flex the patient's thighs on his chest and then extend the legs. This stretches the meninges and, if they are inflamed, causes pain and the patient, in defence, flexes his neck (Fig. 24).

Fig. 24

Doll's eye movements.
Turning patient's head briskly from side to side (but this should not be done in patients with suspected head trauma), the eyes normally move together in the same direction.

This reflex (the so-called oculocephalic response) is absent when there is damage to the brainstem: turning the head, the eyes then move in the opposite direction of the head's movement. (Doll's eye movement).
# NEUROLOGICAL EXAMINATION:
## A SUMMARY

<table>
<thead>
<tr>
<th>Category</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>MENTAL STATUS</strong></td>
<td>Conscious state - behaviour - attention - concentration - language - memory - reasoning</td>
</tr>
</tbody>
</table>
| **MOTOR SYSTEM** | Gait: ataxia - spasticity - parkinsonism  
Balance: Romberg sign  
Involuntary movements:  
thetosis - dystonia - chorea - tremor - myoclonus  
hemiballism - tics - fasciculation  
Limb tone: spasticity - rigidity - flaccidity  
Strength: weakness - paralysis  
Muscle bulk and consistency  
Coordination |
| **CRANIAL NERVES**| Smell: olfactory, 1st  
Vision: optic, 2nd  
Eye movements: oculomotor, 3rd; trochlear, 4th; abducens, 6th  
Corneal reflex: trigeminal, 5th  
Face and taste: facial, 7th  
Hearing: auditory, 8th  
Pharynx: glossopharyngeal, 9th, vagus, 11th  
Tongue: hypoglossal, 12th  
Muscles of neck and shoulder: accessory, 11th |
| **REFLEXES**     | Muscle stretch reflexes, superficial reflexes  
Pathological reflexes: Babinski sign and reflex in coordination test |
| **SENSATION**    | Pin  
Touch  
Vibration  
Position |
| **MENINGES**     | Brudzinski sign  
Kernig sign |
| **AUTONOMIC SYSTEM** | Sphincter function: bladder - anus |
Chapter 2
PATTERNS OF
NEUROLOGICAL IMPAIRMENT

1. Peripheral neuropathy
2. Motor neuritis
3. Muscle weakness without sensory loss
   a. Myasthenia gravis
   b. Muscular dystrophy
   c. Polymyositis
   d. Motor neuron disease
4. Weakness of the face
5. Weakness of one hand and leg
6. Bilateral weakness with sensory loss
   a. Lesion of the neck
   b. Paraplegia
   c. Primary pernicious anemia
   d. Motor neurone disease
7. Disturbance of movement without sensory loss
   a. Tremor and rigidity
8. Ataxia
   a. Drugs
   b. Cerebellar
   c. Due to sensory loss
   d. Familial
9. Athetosis and chorea
10. Sudden paralysis-stroke
11. Deafness and dizziness
12. Sudden transitory loss of consciousness
   a. Syncope
   b. Cardiac
   c. Breath-holding
   d. Transient ischemic attack
   e. Hysteria
   f. Seizure (epilepsy)
   g. Hypoglycemia
   h. Narcolepsy
   i. Heat stroke
13. Head injury
14. Severe headache of rapid onset
15. Blindness
16. Loss of speech


PATTERNS OF NEUROLOGICAL IMPAIRMENT

Injury or disease of various parts of the nervous system produces disturbances of sensation or movement which depend upon which parts of the system are damaged.

A careful neurological examination will reveal the location of the damage. The history will frequently reveal the cause. Sometimes special laboratory examinations are required to verify these clinical findings.

The following sections describe some of the patterns of deficit that may be found on neurological examination and the location and distribution of the causes.

1. NUMBNESS AND WEAKNESS OF HANDS AND FEET

Injury of a nerve produces impairment of sensation and muscle power in the part of the body that nerve supplies.

When the nerves are affected diffusely, the whole body may be involved. The most striking changes are likely to appear first at the ends of the longest nerves - that is, in the hands and the feet. This is termed «polyneuropathy»: patients with this condition complain of weakness, tingling and numbness in the hands and the feet (Fig. 1).

<table>
<thead>
<tr>
<th>MOTOR SYMPTOMS:</th>
</tr>
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<tbody>
<tr>
<td>weakness and final wasting of the muscles (especially distal).</td>
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<tr>
<td>Tendon reflexes are first diminished, then disappear.</td>
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<table>
<thead>
<tr>
<th>SENSORY SYMPTOMS:</th>
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<tbody>
<tr>
<td>sensation of numbness or tingling (paresthesia), then sensory loss in the extremities (pain-touch-pressure-vibratory, or pain-temperature, or all sensations together).</td>
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</tbody>
</table>

Symptoms: the grasp becomes weakened, the legs are unsteady, and the patient cannot rise on his heels or toes. The tendon reflexes are absent. If the condition persists, there is wasting of the muscles in the hands and the feet.

Sensation of pain, temperature, touch, position and vibration are diminished or lost in the extremities.

Causes: there are innumerable causes for these peripheral neuropathies. On a worldwide basis, diabetes and leprosy are the commonest. Chronic alcoholism is also a
common cause. In industrialized countries chemical agents are occasionally responsible. There have been epidemics due to lead, mercury, arsenic, synthetic motor oil (erroneously used as cooking oil), and pesticides. There are also natural plant toxins such as cyanides which produce neuropathy. The occurrence of cases of peripheral neuropathy within the community should lead to careful review of dietary habits and a search for possible exposure to toxins. Cure depends upon correction of deficiency with dietary supplements or upon removal of the toxin.

A rapidly developing and often severe form of neuropathy (Guillain-Barré Syndrome), may result from an unusual response to some common infections or vaccination. Ten days to two weeks after recovering from a flu-like or gastrointestinal illness, the patients develop an ascending weakness which may even progress to complete paralysis. Even the muscles of respiration may be involved so that prolonged periods of assisted ventilation are required. This condition is self-limited and gradual recovery may take place over a period of many weeks. The crucial problem is to maintain the victim by careful nursing through the critical period of respiratory paralysis. Specific treatment (plasma exchange) is available for severe cases.

2. NUMBNESS AND WEAKNESS OF A PART OF THE BODY

Disease, injury, or compression of individual nerves result in loss of sensation and weakness of the muscles in the area supplied by that nerve.

In some conditions, several nerves are involved sequentially.

Causes: inflammatory disease of the blood vessels may be responsible (vasculitis). In some countries, leprosy is the most common cause; in others, diabetes is important. Patients with unexplained focal neuropathy should have appropriate tests for these conditions.

Painful radiculopathy in the upper limbs is often the result of nerve root compression from cervical disc or bony disease. Similarly, painful radiculopathy in the lower limbs causes pain in the back and legs (sciatica) and it is frequently the result of degenerative disease of the lumbar spine.

### PERIPHERAL NEUROPATHIES: CLINICAL SYNDROMES

- **ASSOCIATED WITH SYSTEMIC DISEASE**
  - diabetic neuropathy
  - uremic polyneuropathy - amyloid neuropathy - dysproteinemic neuropathies
- **INFECTIOUS AND POSTINFECTIOUS NEUROPATHIES**
  - herpes zoster - leprosy neuropathy - diphtheritic neuropathy
  - acute idiopathic polynueuritis (Guillain-Barré), Lyme disease
- **ASSOCIATED WITH CONNECTIVE TISSUE DISORDERS**
- **NUTRITIONAL DEFICIENCY**
  - alcoholic neuropathy - beriberi - vitamin B₁₂ and folate deficiency
- **DUE TO COMPRESSION AND ENTRAPMENT**
  - carpal tunnel syndrome - ulnar neuropathy - common peroneal nerve palsy
- **TOXIC NEUROPATHIES**
- **HEREDITARY NEUROPATHIES**
3. MUSCULAR WEAKNESS WITHOUT LOSS OF SENSATION

a) An unusual condition, myasthenia gravis, is characterized by muscle fatigue on sustained activity. It leads to drooping of the eyelids, double vision, weakness of the facial muscles and difficulty in speaking, chewing and swallowing. The arms and legs are also involved, and in some cases the muscle of respiration are weakened and breathing becomes difficult.

The diagnosis may be confirmed by the injection of edrophonium chloride, which causes prompt relief of weakness (Fig. 2).

**TENSILON TEST FOR MYASTHENIA GRAVIS**

1. Intravenous injection of Edrophonium chloride (Tensilon), 2-10 mg. Begin with 2 mg, then proceed slowly with the rest.
2. If positive, the response appears after 10-30 seconds.
3. The improvement of muscle strength lasts about 5 minutes.

Fig. 2

Pyridostigmine bromide and neo-stigmine are effective forms of treatment. Some patients require cortico-steroids, but these drugs may cause a temporary worsening with respiratory paralysis and for this reason steroids may be used only in environments where assisted ventilation is available (hospitals). They are often given in association with immunosuppressant drugs. This again requires careful supervision. Plasma exchange may be also considered in extreme cases.

b) A number of inherited diseases cause degeneration of muscle or "muscular dystrophy" (Fig. 3). In these conditions, the weakness affects the muscles of the trunk, neck, and proximal parts of the legs rather than the distal ones. Some forms affect primarily the shoulder girdle and face; others the lower trunk and thighs. Weakness develops gradually during childhood, adolescence or adult life. There are often affected relatives. There is no specific treatment.

c) Inflammation of the muscles may cause pain and generalized weakness. Muscle inflammation can be caused by trichinosis infection from eating inadequately cooked pork. Another cause is dermatomyositis or polymyositis, a condition due to abnormal immune response of the body. This may require steroid treatment.

d) Motor neuron disease (amyotrophic lateral sclerosis) often begins with weakness and muscle wasting. It may affect the upper and lower limbs or the bulbar (throat) muscles and is a steadily progressive disorder. There is often frequent, spontaneous muscle twitching called fasciculation which is not seen in muscle diseases. The
tendon reflexes are hyperactive and plantar responses abnormal. This condition is distinguishable from peripheral neuropathy in that there is no disturbance of sensation.

In most communities this is a rare condition, but there are areas in the world where it is very prevalent. If the latter is the case, this should be reported, since there is important research attempting to determine the cause of amyotrophic lateral sclerosis in these areas of high prevalence.

<table>
<thead>
<tr>
<th>TYPE</th>
<th>PATTERN</th>
<th>AGE OF ONSET</th>
<th>PROGRESSION</th>
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<tbody>
<tr>
<td>Duchenne's</td>
<td></td>
<td>4 years</td>
<td>rapid, fatal</td>
</tr>
<tr>
<td>Limb-girdle</td>
<td></td>
<td>adolescence</td>
<td>slow</td>
</tr>
<tr>
<td>Facioscapulo humeral</td>
<td></td>
<td>variable</td>
<td>slow</td>
</tr>
<tr>
<td>(Landouzy-Dejerine)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Oculo-pharyngeal</td>
<td></td>
<td>late onset</td>
<td>slow</td>
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<tr>
<td>Myotonic</td>
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<td>variable</td>
<td>slow</td>
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</table>

*Fig. 3 Characteristics of muscular dystrophies*
4. WEAKNESS OF THE FACE

Injury of the facial nerve causes weakness of the forehead, eye and mouth on one side. Taste sensation is lost on the front part of the tongue on that side.

Rapidly developing facial paralysis characterizes Bell's palsy, a condition of unknown causation. There is often aching pain behind the ear and within a few hours the face becomes twisted. This condition is self-limited. Recovery takes place over a period of weeks or months.

Slowly developing facial paralysis may be due to a nearby tumour compressing the course of the nerve.

Facial paralysis due to injury of the facial nerve must be distinguished from weakness of the face due to brain damage:
1) when the nerve is affected, there is weakness of the forehead, eye, and mouth;
2) when the brain is affected above the level of the facial nerve nucleus, only the lower face is weak, and the hand may also be weak.

5. WEAKNESS OF THE LOWER FACE, HAND AND LEG OF ONE SIDE OF THE BODY

If the right side of the body is affected, this condition may also be associated with loss of speech: this pattern of one-sided paralysis suggests disease or injury of the brain on the opposite side. It may result from anything which produces pressure or damage to the brain such as tumour, abscess, or cyst. When this process comes on rapidly, it is probably due to stroke. Slowly developing paralysis of this sort requires a special list of studies as it may be remediable by surgery.

6. WEAKNESS OF BOTH SIDES OF THE BODY

Bilateral weakness usually indicates muscular diseases, neuromuscular transmission defect or impairment of function of the anterior horn cells of the spinal cord. The exact pattern depends upon the nature of the pathological process.

a) Pain, Numbness and Weakness of the Arms and Hands and Stiffness and Weakness of the Legs.

Examination reveals numbness in the shoulder and arm area corresponding to the nerve roots in the neck. The tendon reflexes of the arms are variable - often the biceps reflexes diminish but the triceps and finger reflexes increase.
Tendon reflexes in the legs are increased and the plantar responses are abnormal.

This picture results from damage in the neck region and is common after accidental injury. It may easily be overlooked in patients with head injury, since it is a frequent complication. If progressive, it may indicate arthritis of the spine, tumour, tuberculosis of the vertebrae, and narrowing of the spinal canal. It requires special investigation and frequently surgical treatment.

b) Weakness and Loss of Sensation of Both Legs and Feet.

Involvement of the spinal cord in the trunk region causes loss of sensation and weakness below that level. Sometimes there is a band of numbness around the body. The legs are stiff and weak, the tendon reflexes are overactive and plantar responses are abnormal. Sense of position and vibration are impaired in the feet. Bladder function is often impaired.

Paralysis of this sort is frequent after accidental spinal injury. Gradually progressive symptoms suggest pressure. Especially if there is pain in the back and rapid progression, emergency treatment — often surgical — is required. The process may be due to tuberculosis or other infection pressing on the spinal cord. Surgery treatment may be considered because in many cases permanent paralysis may result if the disease progresses.

c) Numbness of the Hands and Feet with Unsteady Gait, Stiff Legs.

Examination reveals marked loss of sense of vibration and position in the feet with weakness, hyperactive tendon reflexes and abnormal plantar responses.

This pattern may result from pernicious anemia. It may be confirmed by blood test, and it may be cured by injections of vitamin B₁₂.

d) Weakness and Wasting of Hands and/or Feet with Hyperactive Tendon Reflexes and Abnormal Plantar Response but without Pain or Loss of Sensation.

See Motor neuron disease, section 3, d.

7. IMPAIRMENT OF MOVEMENT WITHOUT LOSS OF SENSATION

A number of disorders of the brain lead to loss of proper control of voluntary movements. These are frequently associated with changes in muscle tone and with inability to move smoothly.
a) Tremor and Rigidity

*Parkinsonism* is a condition characterized by rhythmic resting tremor, rigidity, and slowness of movements. There are no sensory disturbances.

It is most common in elderly people and often shows itself initially by impairment of gait.

Afflicted persons walk with slow, shuffling steps and without the usual rhythmic swinging of the arms used to maintain balance, and have difficulty in starting or stopping movement, for example, as in getting up from a chair.

The facial expression is rather characteristic - automatic movements of the smile are lost and the face is expressionless.

There is usually an associated tremor at rest, particularly of the hands: it is described as «pill-rolling movement» (Fig. 4). The thumb and forefinger rhythmically rub against each other about seven times a second.

Rigidity and tremor can be demonstrated by attempting to flex and extend the wrist. There is increased resistance both to flexion and extension throughout the movement, and the rhythmic alteration in tone may be felt as an underlying «cogwheel» sensation. Rapid alternating movements of the fingers are impaired: when individuals are asked to approximate the thumb and forefinger rapidly, they soon lose this ability and the hand closes into a fist. A rapid relief of these parkinsonian symptoms may be achieved by appropriate medication.

**8. UNSTEADY GAIT - ATAXIA**

*Ataxia* results from inability to regulate voluntary movements. When the individual tries to use his hands in reaching or grasping, he cannot direct them smoothly and accurately. Thus the movement is tremulous and irregular and frequently misses the mark. Similarly, the control of movement of the legs is impaired and the sufferer walks...
as if drunk - unable to perform smooth movements of the legs. Muscle tone is generally reduced. The extremities are floppy, but the reflexes are present and are not abnormal.

a) The commonest cause of ataxia is intoxication usually with alcohol or drugs. This ataxia is transitory, wearing off when the drug is eliminated.

However, a long continued use of alcohol may lead to peripheral neuropathy and permanent ataxia.

Epidemics of ataxia have occurred particularly in environments where there were toxins in the diet.

b) One form of ataxia is due to injury or disease of the cerebellum or its pathways in the adjacent brainstem. This ataxia may be unilateral or bilateral. When associated with headache, it strongly suggests the presence of brain tumour which requires surgical treatment.

c) Ataxia may also result from loss of sensation of position and vibration due to involvement of a certain part of the spinal cord or nerves.

Cerebellar ataxia may be distinguished from that due to nerve involvement:

- with cerebellar ataxia, the patient is unable to control his movements whether his eyes are open or closed;
- with neuropathic ataxia, he can correct his movements to some extent by use of visual control, but falls or sways with eye closed (Romberg sign) (Fig. 5).

<table>
<thead>
<tr>
<th>A</th>
<th>CEREBELLAR ATAXIC GAIT = it's produced by cerebellar lesions, and the patient has great instability, even with eyes open.</th>
</tr>
</thead>
<tbody>
<tr>
<td>B</td>
<td>SENSORY ATAXIC GAIT = it's the result of lesions of the spinal cord, roots or peripheral nerves, and is associated with loss of position and vibratory sense in lower extremities and with a Romberg sign.</td>
</tr>
</tbody>
</table>

In addition, the person with neuropathic ataxia has impairment of sensation in the lower extremities. He often complains of numbness of the feet, there is loss of position and vibration senses and the ankle jerks are absent.
An important cause is tabes dorsalis due to syphilis; in this condition the victim also complains of sudden sharp, severe pains extending into the lower extremities. The other causes are those described under neuropathy (section 2).

d) A variety of familial disorders are characterized by ataxia and sometimes they include combinations of ataxia, weakness of the lower extremities and a neuropathy.

Friedreich's ataxia is a familial condition, usually coming on in childhood or adolescence, characterized by weakness and wasting of the legs and ataxia.

The feet develop a characteristic high arch. Curving of the spine (scoliosis) is common. Heart disease is part of this disease and the electrocardiogram may be abnormal. The deep tendon reflexes in the legs are absent, but the plantar responses are abnormal. Sense of position and vibration are impaired in the feet.

Other forms of familial ataxia are due to progressive degeneration of the cerebellum; to pure degeneration of the pyramidal tracts leading to spasticity and overactive reflexes; or to combinations of the above with involvement of the peripheral nerves in the legs.

9. INVOLUNTARY MOVEMENTS

Athetosis and dystonia are conditions characterized by slow, involuntary writhing movements of the trunk and extremities and abnormal postures. They can be caused by brain injury at birth and are also seen in hereditary degenerative disorders. Muscle relaxing agents may provide some partial relief if severe spasms occur.

Choreiform movements occurring with hypotonia and diminished reflexes have many resemblances to ataxia seen in cerebellar disease; however, they are characterized by brief involuntary movements which may appear purposeful in character. The person may make a grimace; thrust out his tongue; move his hand as if to reach for something; or shrug his shoulder. These movements appear voluntary, but the person has no control over them.

Sydenham's chorea, is the result of previous infection with the streptococcus and is characterized by involuntary movements that gradually become severe, affecting motor activities, including arm movements, gait and speech. Patients with this condition, which usually afflicts children, should receive prophylactic penicillin. This is especially important because recurrent infections may lead to irreparable heart valve damage.

A familial form of chorea, Huntington's disease, is an inherited (autosomal dominant) disorder (Fig. 6).
In Huntington's disease, inherited as an autosomal dominant disorder, half of the children of an affected person will develop this condition.

That is, if an individual is affected, one parent has always had this disorder. Half of the children of an affected person will develop this condition which ordinarily comes on during or after adolescence. It is characterized by movements similar to those seen in Sydenham's chorea but it is associated with progressive dementia.

**UNSTEADY GAIT**

(summary)

1. ASSOCIATED WITH SLOW MENTALITY OR CONFUSION
   - Alcohol
   - Other drugs
   - Toxins

2. ASSOCIATED WITH NUMBNESS OF THE FEET AND DIMINISHED REFLEXES
   - Peripheral Neuropathy
   - (unsteadiness made worse by closing eyes)
   - Late Syphilis
   - (if associated with «Lightning Pains»)

3. GENERALIZED INCOORDINATION, ESPECIALLY OF TRUNK AND ARMS
   - Disease of the Cerebellum or Brainstem
   - Hereditary Ataxias
   - Inflammatory or Degenerative Diseases
   - Brain Tumour
10. SUDDEN PARALYSIS OF ONE-HALF OF THE BODY

The most common cause of sudden paralysis is a «cerebral vascular accident». This may be due to blockage of an arteriosclerotic blood vessel in the brain; blockage of a blood vessel by a fragment of a clot (embolus) from some other parts of the circulation - heart or blood vessels; or hemorrhage.

CLASSIFICATION OF STROKES

ISCHEMIC (nonhemorrhagic) PROCESS

- TIA (Transient Ischemic Attack)
- Reversible Ischemic Neurological Deficit (RIND)
- Stroke in evolution
- Cerebral infarction (thrombosis, embolism, others)
- Hypertensive encephalopathy
- Other conditions

HEMORRHAGIC PROCESSES

- Hypertensive intracerebral hemorrhage
- Ruptured saccular aneurysm
- Ruptured arteriovenous malformation
- Hemorrhagic disorders
- Cerebral venous thrombosis

The most sudden episodes are those occasioned by emboli.

A clot lodging in the brain may produce a very sudden paralysis. Within the course of five or ten minutes, the individual may lose speech and become paralyzed on the right side of the body, from a lesion on the left side of the brain, or there may be numbness and paralysis of the left side without loss of speech, if the lesion is on the right side. In instance of embolism, it is important to find the source. Sometimes it is due to infection of the blood stream (septicemia) or chronic heart disease: in these situations, the use of anticoagulants or antibiotic therapy may be life-saving.

Cerebral hemorrhage may also have a rather sudden onset - very commonly with severe headache and paralysis developing over the course of half an hour or one or two hours.

Cerebral hemorrhage is most likely to occur in people suffering from severe hypertension; many cases of cerebral hemorrhage could be prevented by proper control of hyper-
tension. For this reason, everyone should have periodic blood pressure tests, and if the
blood pressure is high, should be given treatment. Measures to maintain a moderate
blood pressure are of extreme importance.

Whereas hypertension is a condition more common in older people, a second cause of
subarachnoid hemorrhage - rupture of an intracranial aneurysm - may occur at any
age. The onset is usually sudden and with a very severe headache, and though there is
frequently mental confusion, the patient is less likely to have a paralysis.

Treatment of this condition requires complete bedrest and maintenance of the blood
pressure at a moderate level by proper medication. However, powerful antihypertensive
agents should not be used because if the blood pressure falls too low, not enough blood
flows to the damaged area of the brain with a worsening of the paralysis. For cerebral
aneurysm, surgical treatment is indicated in many instances.

Cerebral thrombosis has a more gradual onset - sometimes progressing slowly over a
period of hours or even days. The patient may first notice a little weakness of the face or
hand, then this gradually progress to extend more widely, even to complete paralysis of
one side of the body.
<table>
<thead>
<tr>
<th>CHARACTERISTIC</th>
<th>EMBOLUS</th>
<th>THROMBOSIS</th>
<th>HEMORRHAGE</th>
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<tbody>
<tr>
<td></td>
<td>INTRA CEREBRAL HEMORRHAGE</td>
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<td>Anticoagulant</td>
<td>Rest</td>
<td>VASCULAR MALFORMATION AND BLEEDING</td>
</tr>
<tr>
<td>ONSET</td>
<td>Generally during activity</td>
<td>Prodromal episodes of dizziness, aphasia, often with improvement between attacks</td>
<td>Sudden onset of severe headache unrelated to activity</td>
</tr>
<tr>
<td></td>
<td>severe headache</td>
<td>no prodome or amaurosis fugax</td>
<td>no headache</td>
</tr>
<tr>
<td>COURSE</td>
<td>Rapid hemiplegia over minutes to 1 hour</td>
<td>Cerebral progression over minutes to hours</td>
<td>Rapid improvement may occur</td>
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<td>HISTORY AND RELATED DISORDERS</td>
<td>Suspect diagnosis in acute leukemia, aplastic anemia, cirrhosis of the liver</td>
<td>Arterio-sclerosis, diabetes mellitus, xanthomatosis</td>
<td>Emboli in other organs (spleen or kidney) or in several regions of the brain</td>
</tr>
<tr>
<td>SENSORIUM</td>
<td>Coma</td>
<td>Partially preserved</td>
<td>Partially preserved</td>
</tr>
<tr>
<td>NEUROLOGICAL EXAMINATION</td>
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<td>Focal neurological signs</td>
<td>Focal neurological signs</td>
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<td></td>
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<tr>
<td>BLOOD PRESSURE</td>
<td>Arterial hypertension</td>
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<td>Normal</td>
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<tr>
<td>CSF</td>
<td>Grossly bloody</td>
<td>Clear</td>
<td>Clear</td>
</tr>
<tr>
<td>SKULL X-RAY</td>
<td>Shift of pineal to the opposite side</td>
<td>Calcification of internal carotid artery syphon</td>
<td>Pineal ap to show little if any displacement</td>
</tr>
</tbody>
</table>
11. **DEAFNESS AND DIZZINESS**

The symptoms of deafness and dizziness are often associated because the organ for equilibrium (the vestibular organ or labyrinth) is located in the ear.

Dizziness which results from injury or irritation of the internal ear or its connections is true vertigo. It consists of a clear cut sensation of falling or rotation: the person usually states that the world seems to be spinning around. He is often nauseated.

Other forms of dizziness are less clear cut and are more likely to consist of sensations of confusion, faintness, or disorientation. Such symptoms are more commonly attributable to disorders of the cerebellum or other parts of the brain than to the ear. Several patterns of impairment may be recognized.

a) Deafness in one or both ears without dizziness.

Deafness without dizziness is usually attributable to ear disease.

Wax in the ear or infections of the external canal are the most common. Infections of the middle ear are usually associated with cold or sore throat. There is severe ear pain and deafness. Antibiotic treatment is imperative.

In elderly people this may be due to hardening of the bones of the ear (otosclerosis) which convey sound from the eardrum to the sense organ. This condition may be relieved by surgery.

b) Deafness in One Ear with Attacks of Vertigo and Ringing in the Ear (Tinnitus).

This combination of symptoms means injury of the ear or its nerve. Intermittent attacks can be due to inflammation of the internal ear (labyrinthitis).

Slowly progressive one-sided deafness with ringing in the ear may mean a tumour of the acoustic nerve. It can be relieved by surgery and should be treated early. If one-sided or bilateral, it could be due to Ménière's disease.

c) Dizziness without Deafness or Ringing.

Diseases of the labyrinth or of various parts of the brain can produce this symptom. Most commonly, it is the result of a head injury with damage to the labyrinth or to presumed inflammation of this organ (vestibular neuronitis). Both conditions recover spontaneously.

It is also common in elderly persons with cerebral arteriosclerosis. It can be produced by tumours of the cerebellum or upper part of the spinal cord (brainstem). It is a
common symptom in multiple sclerosis: persons with this disease experience recurrent episodes of neurological impairments - first in one part of the body and then another. Common symptoms are dizziness and nystagmus, ataxia with irregular body movements and slurred speech, numbness and clumsiness of the hands, ataxia or stiffness and weakness (spasticity) of the legs.

### DEAFNESS AND DIZZINESS

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<tr>
<th>(summary)</th>
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<tbody>
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<tr>
<td>Wax in the Ears - Ear Infection - Otosclerosis</td>
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<tr>
<td>2. DEAFNESS IN ONE EAR WITH ATTACKS OF DIZZINESS AND RINGING</td>
</tr>
<tr>
<td>Intermittent Attacks = Labyrinthitis</td>
</tr>
<tr>
<td>Slowly Progressive = Tumour of the Acoustic Nerve - Ménière's Disease</td>
</tr>
<tr>
<td>3. DIZZINESS WITHOUT TINNITUS OR DEAFNESS</td>
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<tr>
<td>Injury to Labyrinth - Vestibular Neuritis</td>
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<td>Multiple Sclerosis</td>
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<tr>
<td>Tumors of the Base of the Brain</td>
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<tr>
<td>Parasitic infestation</td>
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</table>

### 12. SUDDEN TRANSIENT LOSS OF CONSCIOUSNESS

Many persons suffer from recurrent episodes of loss of consciousness. They may be difficult to differentiate. Since the doctor rarely sees the episodes, diagnosis depends upon an accurate history.

From the patient, it is important to determine the conditions under which the attack occurred, and especially the feelings or sensations experienced just before loss of consciousness took place. From observers one can determine the mode of onset - the pale color and warmth of the skin, the presence or absence of muscle spasm, the duration of unconsciousness, and the conditions during recovery.

a) Fainting attacks (syncope) are likely to occur after a fright or painful experience. There is a premonition of dim vision, nausea or dizziness. They usually occur when the patient is standing; there is sudden collapse. The hands are cold and sweaty. The pulse is weak. Recovery is rapid when the person is placed in a horizontal position. There may be a brief convulsion at the end of a fainting attack.

b) Irregularities of heart rhythm can cause sudden fainting attacks (syncope). However, they are more common in elderly persons. Frequently the patient is aware of irregular heart beat. Electrocardiographic studies may be required to establish a firm diagnosis.
c) **Breath-holding attacks** - only in infancy. They usually follow a painful experience or a disciplinary measure from a parent. The child screams, then holds his breath and becomes blue. He may continue until there is loss of consciousness. Sometimes there is a brief convulsive spasm.

d) **Transient ischemic attacks** (TIA) occur in elderly people with cerebrovascular disease. They consist of a sudden episode of neurological impairment - numbness or weakness of the face or extremity, partial blindness (amaurosis fugax), dizziness, sudden falling, rarely with momentary loss of consciousness. The patient usually has other evidence of **vascular disease** including abnormal heart sounds and abnormal sounds over the arteries of the neck due to irregularities of the blood vessel wall. **Attacks may herald the occurrence of a stroke** with persistent paralysis.

e) **Anxiety** - Frightened or nervous people may experience «attacks» that resemble loss of consciousness and/or muscle spasm or convulsion. Over-breathing may occur in patients who feel they are suffocating and may even lead to unconsciousness. Attacks usually occur when the patient is nervous and upset or angered and are triggered by circumstances and events in the surroundings. Consciousness appears to be fluctuating and attacks with failing movements or jerking of the trunk may persist from minutes to hours.

f) **Epilepsy** - The attacks of epilepsy are stereotyped and repetitive in character and their onset is unrelated to environmental factors. The onset is sudden, sometimes the patient has some peculiar sensation or feeling (aura) for a few seconds as the attacks begins (see Epilepsy, chapter 6). Jerking spasms may occur - sometimes starting in face, arm or leg, other times affecting the entire body.

g) A rare form of collapse is that associated with unusually low blood sugar (hypoglycemia). It is not uncommon in persons with diabetes from taking **too large a dose of insulin**. Several hours after a meal, the individual becomes weak and tremulous. He has cold sweat and may lapse into confusion or aggression and unconsciousness. He recovers promptly following the administration of sugar.

h) **Narcolepsy** - Narcolepsy is a condition characterized by uncontrollable sleepiness. The patient will drop off to sleep almost instantaneously and unpredictably throughout the day. It is associated with two other symptoms:

- Difficulty going to sleep in the evening because of hallucinations or illusions. As they drift off to sleep, they might have the impression that someone is appearing before them or that people are walking in the room.

- A more important symptom is cataplexy. When people are emotionally stimulated, as by a good joke, as they start to laugh they become extremely weak and fall to the floor. These «catapletic» attacks may resemble seizures.
i) Heat stroke and heat exhaustion - These are two somewhat different reactions to exposure to extreme heat. Persons with heat stroke are very hot, flushed with a high fever and rapid pulse. They respond to application of cold sponges. Persons with heat exhaustion are cold and clammy and have the appearance of syncope. They respond to the administration of salt solution.

<table>
<thead>
<tr>
<th>SUDDEN COLLAPSE WITH LOSS OF CONSCIOUSNESS</th>
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<tbody>
<tr>
<td><strong>HEAT STROKE</strong></td>
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<tr>
<td>PREDOMINANT</td>
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<tr>
<td>AGE</td>
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<tr>
<td>PREDOMINANT</td>
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<td>ONSET</td>
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<td>SKIN</td>
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<td>PULSE</td>
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<tr>
<td>CONVULSION</td>
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<tr>
<td>TREATMENT</td>
</tr>
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13. HEAD INJURY

The most important indicators of the severity of a head injury are

- duration of unconsciousness/duration of post-traumatic amnesia
- presence or evidence of fractured skull
- indications of local brain injury.

It is important at the onset to examine the head with extreme care looking for areas of penetration. One should feel for depressions in the skull from depressed fracture. Evidences of fracture are black and blue areas behind the ears, bleeding from the ears, and «black eyes».

Deepening stupor and inequality of the pupils developing in an individual who has had a recent head injury should immediately lead to a suspicion of hemorrhage. Such
patients should be referred immediately to a centre where surgical treatment is available since trephination is the only measure which is life saving in this situation. Without treatment, death may occur in a matter of hours.

Subdural hematoma produces a very different result. The bleeding is from the veins - not from the arteries, and as a result a clot forms on the surface of the brain. Over a period of weeks or months, this clot expands to produce pressure on a part of the brain. The person gradually develops symptoms of headache, mental confusion, seizures and sometimes weakness of one leg or arm. This condition may occur even after a rather mild head injury and should be suspected when slowly progressive mental symptoms appear. It requires surgical treatment to evacuate the clot.

Persons with head injury frequently have injuries of other parts of the body, especially the neck. For this reason, they must be moved with great caution. They must always be carried with the head directly in line with the body and the head must not be permitted to fall backward or forward or rotate toward either side. Back injury, injury to other parts of the body require concomitant therapy.

Aside from intracranial hemorrhage, a significant proportion of patients with severe head injury develop swelling of the brain and increased intracranial pressure. This condition will also manifest itself with dilated pupils. Patients who have experienced head injury should be on restricted fluid intake until the danger of brain edema has passed. Edema itself is sometimes treated by the use of intravenous hypertonic solutions such as a 20% solution of mannitol.

Dexamethasone is also useful.

14. RAPIDLY DEVELOPING SEVERE HEADACHE

Severe headache may be a dangerous symptom of intracranial bleeding or infection of the brain or meninges; although also seen with a variety of acute fevers, its presence should always raise the possibility of these two conditions, an important clue being the presence of a stiff neck.

Patients suspected of having bleeding or infection should have a lumbar puncture with examination of the spinal fluid for blood, white blood cells and bacteria. Those with hemorrhage should be treated as outlined above with a reduction of blood pressure and sedation. In patients with meningitis, the examination of the skin for small red spots (petechia) may indicate a bloodstream infection. Since the most common variety - Meningococcus meningitis - is epidemic, the presence of other cases in the community may provide a valuable clue. It is an emergency for such patients to receive the appropriate antibiotic therapy.

Recurrent headache is a very common symptom. The most striking form is migraine. Migraine attacks are recurrent, often starting in childhood and being frequently familial.
They are often preceded by disturbances of vision, sensation or even partial paralysis lasting up to 20 minutes before the onset of a one-sided severe pounding headache (classical migraine). There are nausea, vomiting and severe malaise which may last hours and are relieved by sleep.

Migraine attacks may be alleviated by ergotamine, sumatriptan, analgesics preferably given with an anti-ematic, such as metoclopramide, but when frequent (more than twice monthly), can be prevented by the use of propranolol, pizotifen, or flunarizine.

A more usual form is the common migraine attack. These are not preceded by visual phenomena, but consist of a gradually increasing pounding headache. Frequently there is pallor of the face and photophobia.

These attacks are treated in the same way as classical migraine but if depression is associated, amitriptyline is a very effective prophylactic.

A constant dull and progressive headache which is of short duration (usually less than 3 months), worse in the morning and associated with nausea and bradycardia may be an indication of increased intracranial pressure from a brain tumour, abscess, parasitic cyst or a subdural hematoma. The latter may occur days or weeks after a head injury and in an individual who has apparently recovered.

Patients having such headaches should be carefully examined for evidence of neurological impairment, especially minor paresis of the face, arm, or leg. The examination of the eye grounds (optic fundi) is also an important measure in looking for increased intracranial pressure. If these signs are found, referral for neurosurgical study is appropriate.

**Differential Diagnosis**  
RAPIDLY DEVELOPING HEADACHE  
(with stiff neck)

<table>
<thead>
<tr>
<th>CHARACTERISTIC</th>
<th>MENINGITIS</th>
<th>HEMORRHAGE</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>ONSET</strong></td>
<td>Gradual</td>
<td>Sudden</td>
</tr>
<tr>
<td><strong>FEVER</strong></td>
<td>Over 38°C degrees</td>
<td>37-38°C degrees</td>
</tr>
<tr>
<td><strong>RASH</strong></td>
<td>+ or -</td>
<td>No</td>
</tr>
<tr>
<td><strong>PREVIOUS SYMPTOMS</strong></td>
<td>Malaise</td>
<td>Hypertension</td>
</tr>
<tr>
<td><strong>LUMBAR PUNCTURE</strong></td>
<td>White cells</td>
<td>Bloody or hazy</td>
</tr>
<tr>
<td><strong>TREATMENT</strong></td>
<td>Penicillin and chloramphenicol</td>
<td>Bedrest Surgery</td>
</tr>
</tbody>
</table>
Other causes of consistent or recurrent headache include infection of the sinuses or teeth and inflammation of the temporomandibular joint, temporal arthritis and glaucoma.
15. BLINDNESS NOT ASSOCIATED WITH EYE DISEASE

Aside from blindness due to eye disease, there is that resulting from injury to the brain or to the optic nerve and to the tract connecting the eye to the brain.

The connections from each eye meet in the midline (chiasm), then those from the left half of each eye go to the left side of the brain and those from the right half to the right side of the brain. Since the visual image crosses in the retina, this means that the left side of the brain sees only what is on the right half of the vision, and vice versa (Fig. 7).

The pattern of blindness that results from injury depends upon its location and nature.

---

**a) Blindness of One Eye**

Loss of vision in one eye means an injury of the eye or optic nerve in front of the optic chiasm. It may be due to injury of the nerve, tumour or inflammation behind the eye. Sometimes transitory impairment of the circulation to the optic nerve will cause dim vision along with other neurological signs such as aphasia or weakness of the opposite face and hand.
b) Impairment of Vision in Both Eyes

Inflammation of the optic nerve (optic neuritis) may produce loss of vision in one or both eyes. Since the nerves for central vision are the most sensitive, a person may develop a visual loss at the point directly in front of their vision referred to as a scotoma. (Fig. 8).

![Fig. 8](image)

BLIND SPOT

Fig. 8

c) Loss of Vision to One Side

When the injury is behind the chiasm, where the nerve fibers cross, there will be loss of vision to one side in both eyes. Thus, injury of the brain on the left will cause the person to be unable to see anything to the right of the fixation point from either eye (Fig. 9). When this has sudden onset it usually is due to stroke. Gradual loss of vision suggests tumour, abscess or cyst.

![Fig. 9](image)
16. LOSS OF SPEECH (APHASIA)

The centers for speech are located in the left side of the brain, although in some left-handed persons the speech centers are on the right. When these centers are damaged, individuals have impairment in the ability to speak and/or to understand what is spoken to them even though they may be conscious and aware and quite able to move their mouth and tongue.

When the defect is in the front part of the brain near the center for movement, the result is an «expressive or non-fluent aphasia» (Broca's aphasia). The person is able to understand when spoken to but cannot speak. As speech recovers, it is in an abbreviated form – limited to key words in the sentence. The outlook for recovery may be fairly good in this form of aphasia. It is frequently associated with weakness of the face and arm on the right.

When the defect is in the central areas, a more serious deficit occurs as person loss the ability to understand. Sometimes they can repeat what they hear without understanding. Their own speech becomes garbled and incoherent. Sometimes it is like a long string of words that do not make sense. The patient is not aware that what he says is not understandable. This form of speech defect, known as «sensory or fluent aphasia» (Wernicke's aphasia) may occur without any associated paralysis. The outlook for recovery of speech is poor.
Primary preventive neurology
(flow chart No 1)

The potential of the nervous system to recover lost function after injury is less dramatic compared with many other organs. This relatively poor potential for recovery is a consequence of the high level of structural complexity which the human nervous system has attained through the evolutionary process. This complex organization endows humans with tremendous intellectual and motor abilities which should be protected from injury through disease. It is for these reasons that prevention should be emphasized, particularly in neurology, at the primary health care level.

<table>
<thead>
<tr>
<th>RISK FACTOR (to prevent)</th>
<th>CONSEQUENCES</th>
<th>PREVENTIVE MEASURES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prematurity</td>
<td>Cerebral palsy, Epilepsy</td>
<td>1. Good Maternal Nutrition</td>
</tr>
<tr>
<td></td>
<td>Poliomyelitis, Whooping Cough, Tetanus, Measles Encephalopathy, Tuberculous Meningitis (infants), Pyogenic Meningitis (selected group), Diphtheria</td>
<td>2. Avoid Smoking and Alcohol in Pregnancy</td>
</tr>
<tr>
<td></td>
<td>Damage to Brain and/or to Peripheral Nerves</td>
<td>3. Utilize Antenatal Care Facilities</td>
</tr>
<tr>
<td></td>
<td>Damage to Brain and/or to Peripheral Nerves</td>
<td>Immunization Against these conditions: Poliomyelitis, Whooping Cough, Tetanus, Measles, Tuberculosis, Diphtheria</td>
</tr>
<tr>
<td>Infection and inflammation of the nervous system</td>
<td></td>
<td>Avoid: Insecticides, Treated Seed Grain, Paint Spray and Leaded Paint, Leaded Eye Shadow (Sharma), Cleaning Fluids, Contaminated Cooking Oil, Storage Batteries and Battery Cases</td>
</tr>
<tr>
<td>Intoxication of the nervous system</td>
<td></td>
<td>Seat Belt Be Accident Conscious Crash Helmet</td>
</tr>
<tr>
<td>Accidents on highways, at home, at work, at play</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
### Preventive measures in peripheral neuropathies

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Consequences</th>
<th>Preventive Measures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Insulin Dependent Diabetes, Hypoglycemia without warning</td>
<td>Foot cellulitis, sepsisemia, gangrene, amputation</td>
<td>- Strict timing of meals and snacks&lt;br&gt;- Precaution with increased physical activity&lt;br&gt;- Blood glucose monitoring&lt;br&gt;- Training of family and entourage for treatment (carbohydrate, glucagon)</td>
</tr>
<tr>
<td>Infections: infected injuries and plantar ulcers</td>
<td>Bladder: pyelonephritis, loss of kidney function</td>
<td>- Daily hygiene: washing and careful drying&lt;br&gt;- Daily inspection of feet and shoes&lt;br&gt;- Not walking barefoot&lt;br&gt;- Immediate disinfection of cuts&lt;br&gt;- Treatment of interdigital fungus infection&lt;br&gt;- Urination at regular intervals&lt;br&gt;- Manual abdomen pressure&lt;br&gt;- No catheterization</td>
</tr>
<tr>
<td>Infantile Diarrhea</td>
<td>Brain damage</td>
<td>Use latrine, do not wade or swim in contaminated water, cook all meat until brown, periodic stool examination, test and treat pigs&lt;br&gt;Breast feeding, use pure or boiled water</td>
</tr>
</tbody>
</table>
Altered states of consciousness
(the unconscious patient)
(Flow chart No 3)

A systematic approach to etiological diagnosis in patients with altered states of consciousness is necessary if errors are to be avoided or reduced to the barest minimum. Unfortunately, the first contact of the patient with the health care system is often with personnel whose knowledge of the anatomical and physiological basis of the problem is severely limited. Nonetheless, a practical clinical approach at the Primary Health Care level, which emphasizes prevention of irreparable brain damage until support is provided by the higher levels of the health system, can be mastered through an awareness of the relatively small classes of lesions which alter consciousness.

The crucial issues requiring the attention of the primary health worker in such situations are to suspect and detect treatable conditions, be aware that any condition detected may coexist rather than be the primary cause of the patient's illness, and most important, be ready to transfer patients to higher levels of the health system when
diagnosis is in doubt, patient is not improving, or inadequate facilities adversely affect the provision of essential care.

The primary health worker in many developing countries has neither sufficient training nor the authority to prescribe antibiotics, intravenous infusions and other life saving drugs. Therefore, the trained medical personnel at levels of the health system closest to the primary health worker have a vital role to play in the primary care of the unconscious patient. The urgent medical needs of the majority of unconscious patients can be met at the Primary Health Care level, in many developing countries, if available resources are optimally utilized. The following algorithms reveal that only a small proportion of patients with altered states of consciousness need immediate referral to sophisticated medical centres. This latter group includes patients harbouring intracranial mass lesions such as neoplasms, abscesses, hematomas and cerebral edema.

**FLOW CHARTS FOR MANAGEMENT AT PRIMARY HEALTH CARE LEVEL FOR PATIENTS WITH ALTERED STATE OF CONSCIOUSNESS**

1. **COMA**
   - ELEVATED BLOOD PRESSURE?
   - STROKE?
   - PREGNANCY
   - RENAL DISEASE?
   - YES: ? HYPERTENSIVE ENCEPHALOPATHY
   - NO: UNLIKELY HYPERTENSIVE ENCEPHALOPATHY

2. **COMPARE BLOOD PRESSURE:**
   - ISCHEMIC?
   - HEMORRHAGIC
   - CHECK:
     - URINE OUTPUT
     - UREMIC FETOR
     - BLOOD UREA
     - PROTEIN IN URINE

3. **TEST URINE FOR GLUCOSE AND ESTIMATE BLOOD GLUCOSE:**
   - YES: PROBABLY HYPOGLYCEMIC COMA
   - NO: HISTORY OF DIABETES MELLITUS
   - START DEXTROSE INFUSION
   - BOLUS OF CONCENTRATED DEXTROSE 50% - 10%
   - DRAMATIC IMPROVEMENT
   - MD: STOP DEXTROSE INFUSION BUT KEEP SALINE AND REFER MD

4. **CHECK FOR:**
   - ALCOHOL ODOR IN BREATH
   - HEAD INJURY
   - LIVER FAILURE
   - MENINGITIS
   - GIVE FLUIDS, ANTICIPATE WITHDRAWAL SYNDROME

5. **DRUG INTOXICATION?**
   - INTENTIONAL OR ACCIDENTAL POISONING?
   - GASTRIC LAVAGE IF DISCOVERED EARLY (<3 hours) + I.V. FLUIDS, THEN ADDRESS PRIMARY CONDITION
### Examination of the mental state
(Flow chart No 4)

Several treatable medical disorders can impair a patient's mental state. The degree of impairment may vary from lethargy and mild intellectual decline to loss of consciousness. Unfortunately, many patients who present with mild to severe decline in memory and intellectual function have progressive incurable neurological conditions (Alzheimer's disease) rather than treatable medical disorders. However, the search for a treatable cause is worthwhile because very good results can be expected from treatment of conditions such as subdural hematomas, myxedema, nutritional deficiency, pernicious anemia and benign intracranial tumors.

In developing countries, acute devastating illnesses such as childhood diarrheal diseases, infections and infestations of various kinds are of such magnitude that patients with declining intellect, particularly when they are elderly, are often neglected. The needs of such patients are likely to remain unaddressed until improvements in the organization and administration of health services ease the pressure on the limited resources and
permit higher levels of the health system to provide the required support for patients who cannot be managed at the primary health level. A simple screening algorithm is given for patients suspected to be suffering from dementing illnesses.

**ASSESSMENT OF MENTAL STATE**

- **IS THE PATIENT ALERT, AND DOES HE KNOW WHERE HE IS, THE DATE, DAY, MONTH AND YEAR?**
  - **YES**
  - **NO**

- **IS PATIENT'S MEMORY AS GOOD AS IT ALWAYS WAS?**
  - **YES**
  - **NO**

- **DOES THE PATIENT EXPRESS HIM/HERSELF ADEQUATELY AND NORMALLY, AND DOES HE/SHE COMPREHEND SPOKEN AND WRITTEN LANGUAGE?**
  - **YES**
  - **NO**

- **DOES THE PATIENT DRESS AND USE TOILETS APPROPRIATELY?**
  - **YES**
  - **NO**

- **IS THE PATIENT ABLE TO USE MONEY AND GIVE USEFUL ADVICE TO PEOPLE?**
  - **YES**
  - **NO**

- **IS THE PATIENT NOW QUARRELSOME AND SUSPICIOUS?**
  - **YES**
  - **NO**

- **DOES THE PATIENT LOSE HIS WAY AROUND TOWN AND HOME?**
  - **YES**
  - **NO**

- **HAS THE PATIENT DEVELOPED LIMB WEAKNESS, ABNORMAL GAIT, HEADACHES AND VOMITING?**
  - **YES**
  - **NO**

- **IS THE PATIENT NOW UNUSUALLY WITHDRAWN, EXUBERANT, AND DOES HE CRY AND/OR LAUGH EXCESSIVELY AND/OR INAPPROPRIATELY?**
  - **YES**
  - **NO**

- **IS THE PATIENT NO LONGER ABLE TO UNDERSTAND PROVERBS?**
  - **YES**
  - **NO**
Approach to diagnosis of convulsive disorders in children under 6 years of age
(Flow chart No 5)

Convulsive disorders in children under 5-6 years of age may be due to fever, or to epilepsy or other damage to the brain. It is important to distinguish the two diagnoses: convulsions due to fever occur only when fever is present and high, and usually do not need any antiepileptic medical treatment, but the medical approach consists in lowering body temperature with antipyretic drugs. Convulsions not due to fever occur with and without fever, and children need investigation to determine the true cause of fits, and to start with an appropriate antiepileptic treatment. The following flow-chart is a guide to distinguish both cases in children with fits and fever.

Flow chart No 5

Note: repeated febrile convulsions require the prophylactic administration of phenobarbital daily for up to two years of treatment to avoid the development of epilepsy.
### Schema for differential diagnosis between common neurological disorders at a primary health care level

*(Flow chart No 6)*

<table>
<thead>
<tr>
<th>DISEASE</th>
<th>HISTORY</th>
<th>SENSORIUM</th>
<th>MEDICAL EXAMINATION</th>
<th>PUPIL REFLEX</th>
<th>CSF CELLS</th>
</tr>
</thead>
<tbody>
<tr>
<td>HEAD INJURY</td>
<td>Injury followed by immediate unconsciousness or progressive stupor</td>
<td>Depends on the severity of trauma</td>
<td>Headache, vertigo, vomiting, confusion, confusion of the head</td>
<td>Pulse may be slow; normal; body temperature, respiration may be variable</td>
<td>Variable response to light, unilateral dilated pupil suggests need for emergency treatment</td>
</tr>
<tr>
<td>SEIZURES (examine after seizure)</td>
<td>Sudden onset, history of previous attacks</td>
<td>Disturbed consciousness during the attack, amnesia for the attack</td>
<td>Vomiting, nausea, spasm, no headache</td>
<td>Pulse is rapid; temperature is normal, respiration is rapid</td>
<td>Response to light absent during the attack</td>
</tr>
<tr>
<td>CEREBRAL VASCULAR ACCIDENT</td>
<td>May be sudden or gradual</td>
<td>Variable, may rapidly progress to coma</td>
<td>Headache, convulsions, vomiting, paralysis, loss speech</td>
<td>Fever, slow pulse, rapid respiration</td>
<td>Response to light may be absent</td>
</tr>
<tr>
<td>MENINGITIS</td>
<td>Slow onset with headache and fever</td>
<td>Drowsy</td>
<td>Nausea, vomiting, headache, stiff neck, Kerning, Brudzinski, convulsions</td>
<td>Fever, chills, rapid pulse, pink spots on skin</td>
<td>Response to light may be absent</td>
</tr>
</tbody>
</table>
MENTAL STATUS EXAMINATION

To examine the present mental state, one must have information about the past history.

Basic data from the past medical history include answers to questions about birth injuries and developmental disorders, as well as about major medical illnesses, such as meningitis, encephalitis, other serious infectious illnesses, heart disease, high blood pressure, diabetes and anemia. One must also ask about a family history of neurological or psychiatric illnesses. In addition, the examiner should know about any possible exposure to toxins, heavy metals, drugs, or alcohol, and must be aware of the patient's nutritional status.

For each abnormal element of the mental status examination, the examiner should consider its temporal profile. Is the abnormality rapidly or slowly progressive? Intermittent (recurrent or relapsing-remitting) or acute? If it is acute, does it then become stable, does it get worse, or does it improve?

**Fig. 1**
For example, a subacute, progressive deterioration of consciousness might make one think of a cerebral infection or a cerebral metabolic abnormality which could be reversed if treated on time; or, a language disorder of acute onset with rapid recovery might make one think of a stroke.

The following presents the outline for a mental status examination (Fig. 2).

In the section which follows, each of these six elements of the mental status examination will be explained, and examination techniques will be provided. The following indicates which parts of the brain are related to each element of the mental status examination.

1. DISORDERS OF CONSCIOUSNESS

At the beginning of any mental status examination, the level of consciousness of the patient should be determined.

The following is a useful clinical guide to determining the level of consciousness.

If the examiner has difficulty determining the level of consciousness, he or she should note the nature of the stimulus required (normal voice, loud voice, painful stimulation, etc.) and the nature of the patient's response (speaking, groaning, movement, etc.).

Observe the unconscious patient carefully, and the response to noxious stimuli; these evoke the motor response, and may be delivered to the supraorbital ridge, the nail bed, or the sternum.
Position of the unconscious patient

1. The arms are flexed and the legs are extended: it is a decorticate posture. There may be destruction of a cerebral hemisphere or of the central core of the brain (diencephalon) or there may be a toxic or metabolic abnormality (Fig. 3a).

![Decorticate Posturing](image)

Decorticate rigidity: it is a type of reflex posturing in which the arms are flexed and adducted, and the legs are extended. It indicates a hemispheric lesion and, if bilateral, it often represents the first stage in central herniation.

Fig. 3a

2. If the arms and legs are both extended (decerebrate posture), there may be destruction or metabolic abnormality affecting the brain stem (midbrain or pons) (Fig. 3b).

![Decerebrate Posturing](image)

Decerebrate rigidity: in this posturing the patient extends, abducts and internally rotates the arms and extends the legs. There is opisthotonos teeth are clenched. This posturing indicates generally bilateral pyramidal tract dysfunction at the midbrain or pontine level.

Fig. 3b

If the patient seems to be asleep, but the muscles are twitching and the body is jerking, the patient may have a metabolic abnormality (such as lack of oxygen to the brain or elevated nitrogenuous waste products or amines in the blood because the kidneys or the liver are not working properly) or the patient may have epilepsy.

Breathing of the unconscious patient (Fig. 4)

1. If the breathing is a regular pattern of getting faster, getting slower, getting faster, getting slower, stopping, etc. (Cheyne Stokes respiration), that often means that both cerebral hemispheres are impaired and that the brain stem is intact; it may also be seen with metabolic disorders and with severely impaired function of the heart.
2. If the unconscious patient has only a rapid, deep breathing pattern (central neurogenic hyperventilation), that often means that there is a damage in the mid-brain or a toxic or metabolic abnormality or an abnormality related to diabetes.

3. If the patient is unconscious and the breathing pattern is unusually slow, that often means that the patient has had a drug overdose or that the brain stem centers controlling respiration are depressed.

If possible, blood should be drawn and examined for signs of drug overdosage or other toxins, or for metabolic abnormality such as abnormalities of sugar, calcium, nitrogen, or liver function.

**ABNORMAL RESPIRATORY PATTERNS**

- **LESION: HEMISPHERES**
  - Hyperventilation
  - Apnea

- **LESION: M IDBRAIN**
  - Cheyne-Stokes respiration: periods of hyperventilation followed by periods of apnea.

- **LESION: BRAINSTEM**
  - Central neurogenic hyperventilation
  - Apneusis: a prolonged inspiratory cramp (holding of the breath on inspiration)
  - Cluster breathing: clusters of 3-4 breaths followed by short periods of apnea
  - Ataxic breathing: a completely irregular pattern of respiration

**Fig. 4**
Deficit of attention

A deficit of attention can usually be suspected in patients who cannot maintain the usual level of concentration expected during the taking of the medical history. These difficulties result from failure.

1. to maintain concentration
2. to shift concentration appropriately, or
3. to suppress inappropriate distractions.

Observation by the examiner during the general examination will suggest the presence of these deficits. More formally, they can be documented by using the digit span test (normal can repeat 7 digit ± 2), or by asking the patient to list all the letters that rhyme with the word «tree». An acute confusional state is often caused by a treatable metabolic or toxic disorder. Disturbances of memory can be a sign of mental decline, especially in old age. They are also found in nutritional deficiency diseases, such as Korsakoff's syndrome. It is important, however, to realize that not all memory disorders are the result of brain disease. Some depressed patients may seem to have a memory disorder, and may be mistakenly diagnosed as having dementia.

2. DISORDERS OF MEMORY

Memory refers to the process of storing experiences, feelings, and perceptions for recall at a later time. In the clinical examination we consider the time span of the patient's life for labelling the different types of memory.

The examination for memory disorders begins with the history. Occasionally a patient will complain of a memory problem, but usually this will be omitted in the chief complaint. However, the clinician should be alerted by the patient who is a «poor historian».

There are many reasons why a patient will have difficulty in the formulation of an accurate history and memory disorder is one of them.

The examination should then follow in a logical manner (Fig. 5).

**Fig. 5**

| IMMEDIATE MEMORY – usually about 60 seconds |
| RECENT MEMORY – for day-to-day and current events |
| REMOTE MEMORY – for early recollections from the patient's life |
First, alertness and attention must be assessed. At this point a test of digit span (to test immediate memory) is useful.

Second, one should assess retrieval of previously acquired information. This should be done for recently acquired information (recent memory) and then for progressively more remote information. One can start with questions related to events of the day (details of meals, schedule, etc) or current events (political, athletic or in whatever area the patient has some knowledge) and proceed to past personal and past political, athletic or cultural information. Most of this information is usually obtained when one takes the general medical history; however, questions should be asked for which one is certain of the correct response.

Third, the capacity to learn new information: this can be done informally by asking the patient to recall information one has previously discussed. More formally, one can ask the patient to recall a list of three unrelated items in 5-10 minutes. However, one must first be assured that the patient is able to recall the objects immediately.

Post-traumatic amnesia occurs in association with head injury, typically in an injury sufficient to cause a concussion (transient alteration of consciousness associated with closed head injury).

Korsakoff's syndrome is a complex of clinical features manifested by amnesia, and, in many cases, confabulation (the production of bizarre and incorrect statements). Korsakoff's syndrome has been associated with metabolic or nutritional deficits (in particular, the B-complex vitamins), toxins, central nervous system infections, trauma, vascular lesions and surgically induced lesions. Korsakoff's disease or psychosis is the label usually used to describe the alcohol related syndrome of amnesia.

3. DISORDERS OF LANGUAGE - APHASIA

Aphasia is an acquired disorder of language due to brain damage. Aphasic classification is based on the identification of disorders in:

- language production (speech/writing) and
- language comprehension (auditory comprehension/reading).

When combined with the assessment of the ability to repeat and name, a classification system emerges which aids in anatomical localization. Most patients with aphasia have damage in the left cerebral hemisphere (Figs. 6a & 6b).
1. **SPONTANEOUS SPEECH**
   Is it fluent? Is speech effortless with normal melody, rate of production and phrase length? Or is it non-fluent, slow, labored and hesitant?

2. **COMPREHENSION**
   Does the patient understand spoken words? This should be assessed at various categories and levels of complexity (objects, body parts, complex ideas, etc.)

3. **REPETITION**
   Can the patient repeat words and sentences?

4. **NAMING**
   Can the patient name objects when asked?

5. **READING**
   Silently and aloud

6. **WRITING**

   **Fig. 6a**

   **CLASSIFICATION SYSTEM FOR APHASIAS**

   **Fig. 6b**

Common forms of aphasia.

a) **Broca's aphasia** (expressive aphasia, motor aphasia).

Patients with Broca's aphasia generally have non-fluent speech output and poor repetition, but with relative preservation of comprehension. Non-fluent speech is slow, effortful, agrammatic, and often limited to content words (telegrammatic). Writing is generally comparable to speech output; and reading comprehension is usually similar to auditory comprehension. A long-lasting Broca's aphasia is invariably associated with a right hemiplegia, affecting face and right arm most severely, and there is a lesion in the frontal lobe that involves Broca's Area 44 (Fig. 7).
b) Wernicke's aphasia (receptive aphasia, sensory aphasia).

Patients with Wernicke's aphasia have fluent speech output with poor comprehension, poor naming, and poor repetition. Fluent speech is well articulated with phrases of normal length and with normal melody. Output may, however, be contaminated by substitutions of words or parts of words. Reading and writing are usually comparable to the other language deficits. Unlike patients with Broca's aphasia, patients with Wernicke's aphasia rarely have a hemiplegia. The lesion is in the posterior temporal area and contiguous parietal lobe (Figs. 8 & 9).
c) Conduction aphasia

This uncommon type of aphasia is characterized by a prominent deficit in repetition with relatively preserved spontaneous speech and comprehension. Spontaneous speech is fluent, although often contaminated by occasional word or letter substitution and word finding difficulty. Comprehension is fairly good in general conversation but may break down on more demanding formal testing. Repetition is markedly impaired. Writing is usually comparable to the level of spontaneous speech. Reading aloud parallels repetition, and reading comprehension parallels auditory comprehension. Significant hemiparesis is rare, but a right-sided hemianesthesia syndrome is not uncommon. The lesion is in the posterior Sylvian fissure region and extends into the deep white matter in that area (Fig. 10).

![Fig. 10](image)

Conduction Aphasia

Fluent output
May be impaired
Relatively good
Relatively good silently, abnormal aloud
Impaired

---

d) Transcortical motor aphasia (dynamic aphasia)

Transcortical motor aphasia is characterized by a marked reduction in the amount and complexity of spontaneous speech despite retained ability to repeat sentences, to read aloud and to name objects. Reading comprehension and auditory comprehension are relatively preserved. The relative sparing of repetition abilities distinguishes a transcortical motor aphasia from a Broca's aphasia (Fig. 11).

![Fig. 11](image)

Transcortical Motor Aphasia

Impaired output
Impaired
Relatively good
Relatively good
Good
e) Transcortical sensory aphasia

This type of aphasia is characterized by impaired auditory comprehension despite preserved repetition and fluent output. Spontaneous speech is fluent, but often disrupted by word-finding pauses and occasional word or letter substitutions. Reading aloud is possible but reading comprehension is poor. Writing is usually worse than spontaneous speech. Hemiparesis or prominent sensory abnormalities are not common (Fig. 12).

![TRANSCORTICAL SENSORY APHASIA](image)

**Fig. 12**

f) Anomic aphasia (nominal aphasia, amnestic aphasia)

Anomia is the inability to generate names in confrontation tasks and in spontaneous speech. As a symptom, it is common to all forms of aphasia. When anomia becomes the predominant feature in an aphasic disorder then the term anomic aphasia is used. Auditory and reading comprehension, reading aloud and repetition are all relatively normal. Spontaneous speech is fluent and marked by severe word-finding difficulty (Fig. 13).

![ANOMIC APHASIA](image)

**Fig. 13**

g) Global aphasia

All aspects of speech and language are impaired. There is normally an associated hemiplegia and hemianesthesia. Most patients with a global aphasia secondary to a unilateral lesion will retain the ability to communicate non-verbally and to understand a surprising
amount of the information contained in conversational language. Unlike patients with more widespread cerebral damage, they usually learn to manage many of the activities of daily living and can become relatively independent (Fig. 14).

**GLOBAL APHASIA**

![Diagram](image)

*Fig. 14*

h) Isolation aphasia (mixed transcortical aphasia)

In isolation aphasia, meaningful spontaneous speech is scanty or absent and comprehension is severely impaired. The ability to repeat, however, is strikingly spared. The following summarizes a classification scheme for aphasia (Fig. 15).

**FLUENT APHASIA**

![Diagram](image)

*Fig. 15*
4. DISORDER OF SPATIAL ORGANIZATION

On occasion the clinician will be confronted with a patient who displays evidence of spatial disorganization that cannot be explained on the basis of a primary sensory or motor abnormality. These patients may not be able to find their way around their home or the hospital; they may have a particular difficulty in performing common tasks: dressing, eating, using tools, drawing, driving, putting objects together, etc. They may have a disorder in the ability to maintain the appropriate spatial relationship of a set of objects or images.

The importance of the disorder depends on several factors:

- the lifestyle and daily activities which rely on these functions to varying degrees. A carpenter, for example, relies heavily on the ability to arrange objects in an appropriate spatial relationships; a bus driver must keep the bus route in mind;

- the ability of the patient to recognize the existence of an impairment. Frequently, lesions producing such deficits are associated with the inability to appreciate the existence of a problem or the precise nature of the problem (often called unawareness or denial of illness). This combination is not uncommonly seen in patients with lesions of the right hemisphere.

Attentional disorders are of special interest in disorders of spatial organization. Some patients with frontal lobe attentional disorder may seem to fixate on inappropriate stimuli. This results from an inability to maintain attention to the appropriate task and to suppress inappropriate stimuli. For instance, they may begin a task of drawing a picture of a house, start to draw the roof, and then continue by drawing another roof on top of that and so on.

Hemi-attention or neglect may often be quite dramatic and the patient may entirely ignore one side of space. For instance, he or she may read only one half of the page, copy one half of a figure, eat from one half of the plate (Fig. 16).

![Fig. 16]
Although these problems occur with lesions of either hemisphere, it is most common with lesion of the right hemisphere with hemi-inattention or neglect of the left side of space.

Many patients have difficulties referred to as visuo-spatial disorders. They may not be able to organize objects properly in space. For instance, when asked to draw a figure such as a house, the elements (windows, doors, roof, etc.) may be arranged in a haphazard fashion; or when asked to arrange blocks in a simple pattern they may lose the concept of the overall configuration. These patients usually have right parietal lesions.

Examination for disorders of spatial organization includes several elements. First, the clinician must suspect the existence of such a disorder - usually on the basis of the history obtained from the patient and his or her friends. Then an evaluation of primary sensorimotor function, language, memory and attention are necessary. Each may be a contributing factor in the presentation of the patient's problem. Next a series of specific tests may be used.

a. Does the patient ignore one side of space (usually the left side)? One useful test (test of extinction) involves touching both hands or both sides of the patient's face at the same time and asking which side was touched.

b. Is the patient aware of an abnormality or does the patient deny his or her illness? Sometimes the patient seems to ignore a hemiplegia (usually when the left side of the body is involved).

c. Test drawing ability: observe the drawing for signs of neglect of one side of space or signs of disorganization. A good series of drawings to use for these tests start by asking the patient to draw a square, then a house, then a cube or box (with the 3 dimensional component) and finally a clock, with the hands set at 10 minutes past 11.

d. Ask questions about spatial organization: does the patient know how to get home from the examination centre? How to get from one part of the country to another?

e. Observe the ability of the patient to find his/her way around within the examination centre.
5. DISORDERS OF COGNITION IN DEMENTIAS

Dementia is a deterioration or loss of intellectual function, including memory and related cognitive abilities. Points to be emphasized are:

- dementia refers to a deterioration from a prior state
- dementing illnesses may be reversible (nutritional deficiencies) as well as irreversible
- dementing illnesses may affect patients of all ages but most often with the elderly.

The diagnosis of dementia is made on the basis of associated symptomatology and underlying etiologies. This usually results in a long list of disease states that have to be eliminated in a systematic manner. The disease categories cover all possible medical conditions which may affect the brain: degenerative, metabolic, infectious, toxic, endocrine, nutritional, traumatic, neoplastic and vascular. This approach, however, is not always useful to the clinician seeking to assess the symptoms of dementia.

A symptomatic approach considers dementing illness in 2 main categories: cortical and fronto-limbic (or subcortical).

A cortical dementia implies (Fig. 17):

1. Loss of language function, usually presenting as a difficulty recalling names and then proceeding to resemble in some ways an aphasic syndrome.

2. Loss of ability to execute motor tasks on command or spontaneously which can only be explained on the basis of a cognitive deficit (the patient loses the ability to manipulate utensils for eating despite adequate dexterity and appreciation of what is intended).

3. Loss of an ability to grasp the meaning of otherwise perceived stimuli.

4. Memory loss

5. Visuo-spatial disorganization

The subcortical type of dementias refers to patients who have the following:

1. Slowing of thought processes
2. Emotional changes
3. Difficulty in manipulating acquired information
4. Memory deficit characterized as «forgetting to remember»
The example of cortical dementia is Alzheimer's disease. The onset of this disease can be anywhere from middle age to late life. In patients over 65, the term Senile Dementia Alzheimer Type is in current use. The early symptoms may be quite mild and are often masked by a relatively preserved personality in a patient who otherwise appears to be free of any neurological disease. These patients may have difficulties with memory, word finding and calculation abilities.

The relative preservation of personality and absence of other neurological signs frequently results in the recognition of this disorder only at a more advanced stage. The particular pattern of cognitive deficits may be quite variable and may be dependent on such factors as premorbid functioning, interaction with effects of aging on the brain, interaction with reactive psychiatric changes (particularly depression in earlier stages) and a variability in the disease process as it may affect different cortical areas at different rates.

Fronto-limbic dementias or subcortical dementias are represented by a wide range of disease states including Parkinson's disease and Huntington's chorea. The most striking feature of this group is the slowness in functioning and long latency to respond. This slowness characteristically increases with the increasing difficulty of the task required.

The emotional changes can be variable, but as a group these patients frequently appear apathetic with intermittent outburst of irritability. The difficulty in manipulating acquired information can usually be assessed by noting the patient's ability to handle all of
the elements necessary in solving problems. Some patients have difficulties solving problems in arithmetic despite an ability to understand each of the steps required in the solution. Other patients may have particular problems, for example, in organizing the details of how they would change a flat tire. Still others may have particular problems interpreting proverbs in an appropriate abstract manner. These problems are all independent of aphasic difficulties. The memory disorder does not fit the usual pattern of amnestic syndromes described above. With appropriate cures these patients may indeed demonstrate relatively preserved memory function. The etiologies of these disorders are diverse and can best be pursued by a search for associated neurological deficits.

The dementia associated with normal pressure hydrocephalus is an example of an irreversible form of subcortical type of dementia. Normal pressure hydrocephalus is the term used by most physicians to refer to patients with large ventricles without apparent obstruction, with normal cerebrospinal fluid pressure, and a recognizable clinical syndrome that can be reversed with a surgical shunting procedure.

The clinical syndrome consists of 3 principal features:
- a prominent gait disorder
- an urgency type of urinary incontinence
- a mild to moderate subcortical type of dementia.

Multi-infarct dementia is a form of dementia found in patients with multiple, small, subcortical, white and grey matter infarctions. A history of step-wise progression of neurological illness together with evidence of multifocal neurological signs often helps distinguish this form of dementia from others.

Further evaluation of demented patients can include:
- careful medical evaluation for etiological factors
- laboratory evaluation for metabolic errors
- an electroencephalogram to look for evidence of diffuse or focal disease
- a cerebrospinal fluid examination for evidence of chronic meningitis, including syphilis in selected cases
- a computerized tomographic (CT) scan or magnetic resonance imaging (MRI) of the head to demonstrate the presence of structural changes. A careful neuropsychological evaluation is always useful to document both the degree and the pattern of deficits. Treatment is based on the underlying etiology.
- Hachinski's Scale can help in differentiating Alzheimer's Disease from Vascular Dementia
6. DISORDERS OF PERSONALITY, EMOTION AND THOUGHT CONTENT

Psychiatric syndromes related to neurological disease present the examiner with a challenge that often requires the skills of a specialist in both neurology and psychiatry. These syndromes are frequently marked by changes in behavior, mood, and personality. Distinguishing between the changes in emotion due to neurological disease and those due to psychiatric disease, when they co-exist, is difficult indeed.

The clinician needs a systematic method to evaluate behavioral changes related to neurological disease. In analyzing affect, one must consider

- affective state
- range of affect
- activity level
- rate of change
- relation of affect to mood.

Psychomotor retardation, apathy and indifference may be related to disease in the frontal lobes or the fronto-subcortical system. True depression is a common feature of Parkinson's disease. Euphoria, even in the presence of severe neurological disability is occasionally found in multiple sclerosis. A clear dissociation between mood and affect is a feature of pseudobulbar palsy, due to bilateral frontal lobe pathology.

Occasionally, a clinical picture of dementia may be caused by depression. This has been called «depressive pseudodementia». The clinical features of depressive pseudodementia often resemble those of subcortical dementia. It is necessary to recognize a depressive pseudodementia, because this is a form of cognitive deterioration which can be successfully treated. Paranoia and hallucinations may result from brain lesions. Olfactory, auditory and visual hallucinations have all been described with temporal lobe epilepsy. Transient paranoid delusions may be seen with toxic, infectious, or metabolic disorders of brain function. More fixed forms of paranoia are frequently seen in elderly persons with hearing disability or with sensory deprivation due to reduced vision and hearing.

Disorders of specific brain regions
Localization of function to specific regions of the brain is a complex task. Nevertheless, specific clinico-anatomical correlations have been accepted by many clinicians, and it may be useful to review these.

Frontal lobe
Left frontal lesions are frequently associated with language disorders that vary according to the size and location of the lesion. Elements of Broca's aphasia or transcortical motor aphasia may be present. Lesions of the frontal lobes can also result in disorders of
attention, either generalized or of the hemi-inattention type. Disorders of memory are difficult to assess because of attentional disorders, but a subcortical or fronto-limbic type of dementia may develop, marked by slowness, apathy, concreteness, personality changes and forgetfulness. Perseveration is common in frontal lobe disease.

Personality changes are frequently present with frontal lobe damage and may be characterized by disorders in the motor control of emotion (pseudobulbar palsy) especially in bifrontal diseases; difficulty translating emotion into a facial expression, especially in right frontal lesion; disorders in the comprehension of emotional intention (particularly with right hemisphere lesion); and by true disorders of mood. Oftentimes these patients have a shallow affect with elements of euphoria, irritability and apathy.

Important objective signs of frontal lobe disease are the «forced grasp» and the «snout» reflexes. Forced grasp is elicited by placing the hand in the patient’s palm. A grasping movement results, as if shaking hands. The snout reflex is elicited by stroking the face at the corner of the mouth. The reflex is a pursing of the lips.

Parietal lobes
Unilateral right parietal lobe lesions have been associated with acute confusional syndromes. Either parietal lobe has been associated with hemi-attention syndromes, although these syndromes are more severe following right parietal damage. Language disorders are frequently encountered with left parietal lesions. Visuo-spatial disorders are commonly associated with right parietal lobe lesions, but the left parietal lobe still plays a role in certain aspects of visuo-spatial organization. Deficits associated with parietal lobe lesions are frequently encountered in the cortical dementias.

Temporal lobes
Bilateral temporal lobe lesions, especially those involving inferior and medial temporal structures, have been associated with acute confusional syndromes. Left posterior temporal lobe lesions are frequently associated with aphasic difficulties which vary according to the precise localization. Bilateral lesions of the auditory cortex can result in
a deficit in the non aphasic interpretation of acoustic information (auditory agnosia) which may be most prominent for linguistic information (pure word deafness) or for environmental sounds (auditory sound agnosia or agnosia for environmental sound). Medial temporal lesions can be associated with an amnestic syndrome which may be specific for verbal information in left-sided lesions or for spatial information in right-sided lesions. Elements of temporal lobe lesions are frequently seen in cortical types of dementias, especially Pick's disease. The relationship of temporal lobe lesion to personality disorders is quite complex and is probably related to limbic connections.

Right hemisphere syndrome
Usually associated with a left hemiplegia, the right hemisphere syndrome consists of disorders of spatial organization (manifested in several areas: constructions, drawings, written language, spatial memory and topographical orientation), together with hemi-inattention to the left side of space, unconcern or denial of illness, and impersistence.

Left hemisphere syndrome
Usually associated with a right hemiplegia, the left hemisphere syndrome consists of disorders of language (aphasias) and of disorders of control of voluntary motor behavior (apraxias), together with disturbances in verbal memory.
Chapter 5

BEDSIDE GUIDE TO THE MANAGEMENT OF ALTERED CONSCIOUSNESS (the unconscious patient)

Altered states of consciousness are variably described using such terms as coma, obtundation, stupor, confusion, and semi-coma. These states are comprised primarily of two components:

- arousal, and
- content of consciousness, and these may be differentially affected.

In the assessment and management of patients suffering from these states, the cardinal principle should be that altered consciousness is a SYMPTOM of underlying brain disease and is not a primary diagnosis.

Information must be gathered from the history, physical and neurological examination and laboratory which give clues to the underlying diagnosis. This manual outlines the most important and common cause of altered consciousness to aid rapid diagnosis and appropriate therapy (Fig. 1).

Fig. 1
Imperative to the understanding of altered consciousness is the mastery of the relatively small classes of lesions which may alter consciousness. These are summarized in the following table (Fig. 2):

<table>
<thead>
<tr>
<th>LESIONS THAT MAY ALTER CONSCIOUSNESS</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. DIFFUSE BILATERAL CEREBRAL DYSFUNCTION</td>
</tr>
<tr>
<td>(examples: structural lesions producing herniation, hepatic coma, hypoglycemia, diabetic coma, meningitis)</td>
</tr>
<tr>
<td>2. EXTENSIVE DESTRUCTION OF THE BRAINSTEM ASCENDING RETICULAR ACTIVATING SYSTEMS</td>
</tr>
<tr>
<td>(example: pontine hemorrhage)</td>
</tr>
<tr>
<td>3. PSYCHOGENIC COMA</td>
</tr>
<tr>
<td>(examples: hysteria, catatonic schizophrenia)</td>
</tr>
<tr>
<td>4. DEPRESSION OF BRAIN STEM FUNCTION</td>
</tr>
<tr>
<td>(example: barbiturate poisoning)</td>
</tr>
</tbody>
</table>

Differentiation among these possible etiologies requires a thorough history and neurological examination. One must keep in mind the limited possibilities for etiology of coma and proceed rapidly through a differential diagnosis.

The initial approach to the unresponsive patient

1. Rapid general medical examination to assess vital signs
2. Obtain a history from any relative or friend of patient who is available
3. General medical examination
4. Neurological examination
5. Then, rapidly, generate a differential diagnosis, order the appropriate laboratory studies, and initiate therapy. While waiting for the laboratory tests, a first good thing to do is to inject glucose intravenously. If the unconsciousness is due to hypoglycemia, the patient will wake-up immediately.
GENERAL SCHEMA FOR GENERATING APPROPRIATE NEUROLOGIC DIFFERENTIAL DIAGNOSIS

**COMA**

NEUROLOGICAL EXAMINATION

<table>
<thead>
<tr>
<th>FOCAL</th>
<th>NON FOCAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>TRAUMATIC</td>
<td>ATRAUMATIC</td>
</tr>
<tr>
<td>Subdural hematoma</td>
<td>Cerebral mass lesion with brain stem compression</td>
</tr>
<tr>
<td>Epidural hematoma</td>
<td>Cerebral mass lesion with brain stem compression</td>
</tr>
<tr>
<td>Contusion</td>
<td>Brainstem infarction or hemorrhage</td>
</tr>
<tr>
<td>Subarachnoid hemorrhage</td>
<td>Subdural hematoma</td>
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</tr>
</tbody>
</table>

*Mass lesion include hematoma, hemorrhage, tumor, abscess or swollen, infarcted brain.*
SOME DIFFUSE, MULTIFOCAL OR METABOLIC CAUSES OF DELIRIUM, STUPOR AND COMA

A. Deprivation of oxygen, substrate or metabolic cofactor

1. **Hypoxia** (interference with oxygen supply to the entire brain-cerebral blood flow, CBF, normal)
   a. Decreased blood pO$_2$ and O$_2$ content
      - Pulmonary disease
      - Alveolar hypoventilation
      - Decreased atmospheric oxygen tension
   b. Decreased blood O$_2$, pO$_2$ normal («anemic anoxia»)
      - Anemia
      - Carbon monoxide poisoning
      - Methemoglobinemia

2. **Ischemia** (diffuse or widespread multifocal interference with blood supply to the brain)
   a. Decreased CBF resulting from decreased cardiac output
      - Strokes-Adams
      - Cardiac arrest
      - Cardiac arrhythmias
      - Myocardial infarction
      - Congestive heart failure
      - Aortic stenosis
      - Pulmonary infarction
   b. Decreased CBF resulting from decreased peripheral resistance in systemic circulation
      - Syncope
      - Carotid sinus hypersensitivity
      - Low blood volume
   c. Decreased CBF associated with generalized or multifocal increased vascular resistance
      - Hyperventilation syndrome
      - Hyperviscosity (polycythemia, cryoglobulinemia, macroglobulinemia, sickle cell anemia)
      - Subarachnoid hemorrhage
Bacterial meningitis
Hypertensive encephalopathy
d. Decreased CBF owing to widespread small vessel occlusion
Disseminated intravascular coagulation
Systemic lupus erythematosus
Subacute bacterial endocarditis
Fat embolism
Cerebral malaria
Cardiopulmonary by-pass

3. Hypoglycemia

Resulting from exogenous insulin
Spontaneous (endogenous insulin, insulinoma, liver disease, etc.)

4. Cofactor deficiency

Thiamine (Wernicke's encephalopathy)
Niacin
Pyridoxine
Cyanocobalamin
Folic acid

B. Disease of organs other than brain

1. Disease of nonendocrine organs
   Liver (hepatic coma)
   Kidney (uremic coma)
   Lung (CO₂ narcosis)
   Pancreas (exocrine pancreatic encephalopathy)

2. Hyper- and/or hypofunction of endocrine organs
   Pituitary
   Thyroid (myxedema; thyrotoxicosis)
   Parathyroid (hypo- and hyperparathyroidism)
   Adrenal (Addison's; Cushing's; pheochromocytoma)
   Pancreas (diabetes; hypoglycemia)

3. Other systemic diseases
   Cancer
   Porphyria
   Sepsis
C. Exogenous poisons
   1. Sedative drugs
      Barbiturates and non-barbiturate hypnotics
      Tranquilizers
      Bromides
      Ethanol
      Opiates

   2. Acid poisons or poisons with acid breakdown products
      Paraldehyde
      Methyl alcohol
      Ethylene glycol
      Ammonium chloride

   3. Psychotropic drugs
      Tricyclic antidepressants and anticholinergic drugs
      Amphetamines
      Lithium
      Phenylcyclidine
      Phenothiazines
      Lysergic acid diethylamide LSD
      Mescaline
      Monoamine oxidase inhibitors

   4. Others
      Penicillin
      Anticonvulsants
      Steroids
      Cardiac glycosides
      Cimetidine
      Heavy metals
      Organic phosphates
      Cyanide
      Salicylate

D. Abnormalities of ionic or acid-base environment of central nervous system (CNS)
   Water and sodium (hyper- and hyponatremia)
   Acidosis (metabolic and respiratory)
   Alkalosis (metabolic and respiratory)
Magnesium (hyper- and hypomagnesemia)
Calcium (hyper- and hypocalcemia)
Phosphorus (hypophosphatemia)

E. Disordered temperature regulation

Hypothermia
Heat stroke
Fever

F. Infection or inflammation of CNS

Leptomenigitis
Encephalitis
Acute «toxic» encephalopathy
Parainfectious encephalomyelitis
Cerebral vasculitis
Subarachnoid hemorrhage

G. Primary neuronal or glial disorders

Creutzfeldt-Jacob disease
Marchiafava-Bignami disease
Adrenoleukodystrophy
Gliomatosis cerebri
Progressive multifocal leukoencephalopathy

H. Miscellaneous disorders

Seizures and postictal states
Concussion
Acute delirious states:
- sedative drug withdrawal
- «postoperative» delirium
- ICU psychosis
- drug intoxication

CLUES IN THE ASSESSMENT OF A COMATOSE PATIENT

1. Rapid general medical examination, with particular attention to the following signs (Fig. 3):

   a) BLOOD PRESSURE
   (is there cerebrohypoperfusion secondary to hypotension?)

   b) PULSE
   (is there cerebrohypoperfusion secondary to bradycardia?)

   c) RESPIRATION
   (is there sufficient hypoventilation to produce hypoxia?)

   d) EVIDENCE OF TRAUMA
   (is there an intracranial hematoma underlying outward signs of trauma?)

   e) PUPILARY REFLEXES
   (the single most important variable for assessing presence or absence of brainstem destruction)

   f) ABNORMAL POSTURING
   (is there herniation?)

Fig. 1

Items a, b, and c should be rapidly normalized with pharmacological or mechanical intervention, such as vasopressors and/or mechanical ventilation.

If there is abnormal posturing or grossly absent or asymmetric pupillary reflexes, assume that the diagnosis is a brainstem destruction due to possible herniation and rapidly proceed with:

1. Passive hyperventilation
2. Osmotic agents (intravenous mannitol 0.5 to 1.0 g/kg)
3. Surgical or radiological confirmation of the diagnosis

If there is trauma, the possibility of subdural or epidural hematoma is high and emergency surgical consultation should be sought.

2. Obtain a history from any available relative or friend with particular attention to:

   a) nature of onset of illness
   - sudden, profound deficits are typical for vascular lesions such as cerebral hemorrhage, brainstem stroke, or posttraumatic hematoma.
gradual, progressive deficits are more characteristics of expanding intracranial masses.

intermittent positive phenomena such as jerking of the extremities suggest seizure which may be caused by structural or metabolic abnormalities.

fever suggests infection

b) recent complaints

- headache
- nausea
- vomiting
- focal weakness or loss of sensation, or
- vertigo

all suggest primary intracranial pathology

c) previous medical illness especially:

- diabetes
- liver disease
- use of blood thinners (anticoagulants)
- bleeding tendency
- kidney disease
- heart disease
- seizure disorder
- depression
- suicide gestures
- psychiatric illness
- thought disorders
- alcohol or drug use
- current medications

d) previous neurological illness

- seizure disorders
- brain tumor
- head injury
- syncope

e) previous psychiatric illness

- depression
- hysteria
- schizophrenia
3. General medical examination, with particular attention to:

a) fever (suggesting infection)

b) stigmata of medical illness
   - liver disease
   - alcohol on breath
   - fruity smell of acetone on breath
   - obesity
   - cyanosis

c) neck for rigidity
   - meningitis

d) evidence of toxic ingestion
   - sedative drugs near where patient was found
   - suicide note
   - needle tracks

e) abnormal movements
   - posturing movements of herniation
   - intermittent rhythmic movements
   - multifocal myoclonus to suggest uremia

4. Neurological examination with particular attention to:

a) level of consciousness

b) language function
   - if awake, language difficulty suggests dominant hemisphere lesion
   - scanning speech suggests cerebellar or brainstem lesion

c) eye signs (Figs 4-7):
   - funduscopy (hemorrhages, exudates, papilledema to suggest increased intracranial pressure)
   - pupillary reflexes
   - oculocephalic (doll's eyes) responses
   - oculovestibular (caloric) responses
   - corneal reflexes
   together, they give information about brainstem function from midbrain to upper medulla.
d) other brainstem reflexes
- gag reflex
- spontaneous respiration (if present, medulla is preserved)

e) sensory / motor responses
- follow commands?
- respond to pain?
- localizes painful stimulus and avoids?
- localizes painful stimulus and withdraws asymmetrically?
- respond to pain with abnormal flexion response?
- respond to pain with abnormal extensor response?
(from the first to the last question, there is a progressively lower level of response)

f) tendon reflexes
The preservation of normal pupillary and extraocular reflexes is the most reliable criterion for excluding brainstem destruction as the etiology for coma. Absence of oculocephalic responses or doll's eyes but preservation of normal pupillary light reflex suggests primary brainstem disfunction, particularly from drug intoxication such as barbiturate.

These signs in the presence of symmetrical sensory/motor responses constitute a nonfocal disturbance in a comatose patient. If alert enough to speak, language function must be normal as well in order to define a nonfocal process. The differential diagnosis is as previously described.

<table>
<thead>
<tr>
<th>OCULOCEPHALIC REFLEX</th>
</tr>
</thead>
<tbody>
<tr>
<td>Place patient's head at 30° above horizontal, and quickly flex the head or rotate it to either side. With normal brainstem function, rotation induces lateral eye movement; extension of the neck produces conjugate deviation of the eyes in the downward position, and flexion of the neck, in the upward position.</td>
</tr>
</tbody>
</table>

Fig. 4
**OCULOVESTIBULAR REFLEX**

Caloric stimulation: 10 ml of cold water are injected into the ear canal. If the brainstem is intact, the eyes will slowly turn towards the stimulus. Bilateral cold water against the tympanic membrane produces conjugate downward deviation of the eyes, whereas hot water upward.

*Fig. 5*

**OCULOCEPHALIC REFLEX (doll's eye)**

Oculocephalic stimulation deviates the appropriate eye laterally, and brings the eye which would normally deviate medially only to midline, because of the lesion of the nervous connections. In this case, vertical eye movements often remain intact.

*Fig. 6*

**OCULOVESTIBULAR REFLEX**

With caloric stimulation, the patient shows the same response: only one eye deviates in turn, while the other is on the midline, also with caloric stimulation, vertical eye movements remain often unchanged.

*Fig. 7*
LABORATORY PROFILE FOR THE COMATOSE PATIENT

1. Mandatory, emergency screening tests
   - glucose
   - sodium
   - potassium
   - bicarbonate
   - calcium
   - urea
   - nitrogen
   - osmolality = 2(Na) + (glucose)/18 + (BUN)/2.8
   - arterial pH, pO₂, pCO₂
   - white blood cell count
   - platelet count
   - prothrombin and partial thromboplastin times

2. Mandatory emergency tests if meningitis or encephalitis has not been clinically excluded
   - Cerebrospinal fluid examination, including
     - glucose
     - cell count and differential
     - Gram stain
     - India ink stain if immunosuppressed patient
     - culture established for bacteria, fungi, tuberculosis (TB)

3. Tests which should be sent routinely but do not critically affect immediate diagnosis or therapy
   - toxic screen
   - alcohol level
   - thyroid function tests
   - serum cortisol

In the management of altered consciousness, always think of the most life-threatening possibilities as well as the most reversible. Most acute coma of unknown cause in developed or semideveloped countries is due to either self-induced drug poisoning (including alcohol) or trauma.

GENERAL THERAPEUTIC PRINCIPLES

1. Support the vital signs
2. Give dextrose, thiamine and naloxone to all comatose patients
3. Recognize herniation syndromes, and
4. Decompress central structures promptly
5. Overwhelming infections should be treated promptly and with broad coverage pending precise microbiologic diagnosis.
Once the vital signs are stabilized, direct and streamline therapy to fit the patient:

1. Withdraw any measures unlikely to contribute positively to management.

2. Adjust antibiotics to microbial sensitivities when available. Be sure certain agents have adequate penetration into the nervous system if meningitis or encephalitis is suspected.

**CLUES TO SPECIFIC DRUGS FREQUENTLY CAUSING DELIRIUM, STUPOR OR COMA**

<table>
<thead>
<tr>
<th>DRUG</th>
<th>CHEMICAL DIAGNOSIS</th>
<th>BEHAVIOR</th>
<th>PHYSICAL SIGNS</th>
</tr>
</thead>
<tbody>
<tr>
<td>amphetamine</td>
<td>blood or urine</td>
<td>hypertension, aggressive, paranoid, auditory and visual hallucinations</td>
<td>hyperthermia, hypertension, tachycardia, arrhythmia, pupils dilated, tremor, dystonia, seizures</td>
</tr>
<tr>
<td>cocaine</td>
<td>none available</td>
<td>as above but less paranoid</td>
<td>as above</td>
</tr>
<tr>
<td>psychedelics (LSD, PCP, mescaline)</td>
<td>blood or urine</td>
<td>confused, disoriented, perceptual distortion, distractable, withdrawn or eruptive</td>
<td>hypertension, tachycardia, pupils dilated, nystagmus, hyperactive, dystonia, myoclonus</td>
</tr>
<tr>
<td>atropine, scopolamine</td>
<td>none available</td>
<td>delirium, agitation, visual hallucinations, drowsiness</td>
<td>fever, flushed face, dilated pupils, tachycardia, hot dry skin</td>
</tr>
<tr>
<td>tricyclic antidepressants</td>
<td>blood or urine</td>
<td>drowsiness, delirium and agitation, rarely coma</td>
<td>fever, supraventricular tachycardia, conduction defects, ventricular tachycardia, hypotension, dystonia</td>
</tr>
<tr>
<td>phenothiazines</td>
<td>blood</td>
<td>somnolence</td>
<td>arrhythmias, hypotension, dystonia, fever rigidity</td>
</tr>
<tr>
<td>lithium</td>
<td>blood</td>
<td>lethargic, confusion, mutism, multifocal seizures</td>
<td>distractable, roving conjugate eye movements, pupils intact, tremor, akathisia, paranoia</td>
</tr>
<tr>
<td>benzodiazepines</td>
<td>blood or urine</td>
<td>stupor</td>
<td>respiratory depression may resemble barbiturate intoxication; if severe increased reflexes, myoclonus, dystonia, seizures, tachycardia, heart failure</td>
</tr>
<tr>
<td>methaqualone</td>
<td>blood or urine</td>
<td>hallucinations, agitation</td>
<td></td>
</tr>
<tr>
<td>barbiturate</td>
<td>blood or urine</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>blood or breath</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alcohol</td>
<td>Blood or urine</td>
<td></td>
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<td>---------</td>
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<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>hypothermia, cool dry skin, pupils reactive, dough's eyes absent, hyporeflexia, hypotension, apnea</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>hypothermia, skin cold, pupils reactive, tachycardia</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>needle marks, hypothermia, skin cool and moist, pupils asymmetricaly pinpoint reactive, bradycardia hypotension, hypoventilation, pulmonary edema</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

# CEREBROSPINAL FLUID FORMULA

<table>
<thead>
<tr>
<th>PURULENT PROFILE</th>
<th>LYMPHOCYTIC LOW-GLUCOSE PROFILE</th>
<th>LYMPHOCYTIC NORMAL-GLUCOSE PROFILE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Elevated WBC, predominantly polymorphonuclear leukocytes, low sugar, high protein</td>
<td>Elevated WBC, predominantly lymphocytes, low sugar, high protein</td>
<td>Elevated WBC, predominantly lymphocytes, normal sugar, high protein</td>
</tr>
</tbody>
</table>

**- INFECTIOUS**
- Bacterial meningitis
- Early viral meningitis
- Embolic cerebral infarction with endocarditis
- Parameningeal infection (subdural empyema, brain abscess, cortical vein thrombophlebitis)
- Early tuberculous meningitis
- Acute hemorrhagic leukoencephalitis

**- NON INFECTIOUS**
- Chemical meningitis (contrast media, foreign bodies)
- Behçet's disease
- Mollaret's recurrent meningitis

- Tuberculous meningitis
- Fungal meningitis
- Resolving or partially treated bacterial meningitis
- Viral meningitis

- Viral meningitis
- Viral encephalitis
- Parameningeal infections (mastoiditis, sinusitis)
- Early fungal or tuberculous meningitis
- Parasitic infection
- Polyradiculitis (Guillain-Barré)

- Carcinoma meningitis
- Neurosarcoidosis

- Postinfectious encephalomyelitis
- Active demyelinating disease

---

# ANTIBIOTIC THERAPY OF BACTERIAL MENINGITIS

## OF KNOWN ETIOLOGY

<table>
<thead>
<tr>
<th>ORGANISM</th>
<th>DRUG OF CHOICE</th>
<th>ALTERNATIVE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diplococcus</td>
<td>Penicillin G</td>
<td>Chloramphenicol</td>
</tr>
<tr>
<td>Streptococcus, groups A, B</td>
<td>Penicillin G</td>
<td>Erythromycin</td>
</tr>
<tr>
<td>Streptococcus, group D</td>
<td>Penicillin G and gentamicin</td>
<td>Vancomycin</td>
</tr>
<tr>
<td>Staphylococcus</td>
<td>Oxacillin</td>
<td>Vancomycin</td>
</tr>
<tr>
<td>Listeria monocytogenes</td>
<td>Penicillin G or ampicillin</td>
<td>Erythromycin</td>
</tr>
<tr>
<td>Meningococcus</td>
<td>Penicillin G</td>
<td>Chloramphenicol</td>
</tr>
<tr>
<td>Hemophilus influenzae</td>
<td>Ampicillin or chloramphenicol</td>
<td>Chloramphenicol</td>
</tr>
<tr>
<td>Pseudomonas</td>
<td>Carbenicillin and gentamicin</td>
<td>Polymyxin B, IM and IT</td>
</tr>
<tr>
<td>E. Coli, Proteus or Klebsiella</td>
<td>Chloramphenicol</td>
<td>Gentamicin IV and IT</td>
</tr>
</tbody>
</table>

Chapter 6

EPILEPSY

INTRODUCTION

A convulsion is one of the most terrifying things one can witness. There is no wonder that in many parts of the world it is believed to be a visitation of the devil, and victims are shunned.

An apparently normal person suddenly stops, may scream, falls unconscious, and has massive muscular contractions with salivation, tongue biting, and often incontinence.

This is called a generalized tonic-clonic seizure, and is what one thinks of when the diagnosis of epilepsy is mentioned.

The attacks are due to a sudden excessive electrical discharge in the brain: it starts in one part, and spreads to the rest of the brain.

As well as the convulsion, there are different types of seizures depending on whether the electrical discharge remains local, or spreads to other parts of the brain.

The seizure may be minute, such as an absence attack which lasts only for a few seconds, and often go unrecognized.

It may be a sudden onset of automatic behavior when the patient does not fall, appears to be conscious, but is going through bizarre activities, or experiencing unusual sensations. These have been called psychomotor seizures.

Although a great deal is known about the chemical and electrical changes that take place during a seizure, there is still much to learn about what actually starts a seizure and how it spreads. What is known has led to the development of drugs that can prevent seizures and enable the patient to live a normal life.
TYPES OF SEIZURES

There are many factors which determine the type of seizure. The first is the region of the brain from which the seizure originates. This depends on the abnormal condition in that region: it may be a tumor, inflammation, vascular occlusion, etc (Figs 1 & 2).

Another factor is the degree of maturation of the brain: a newborn may have migrating focal motor seizures, while the infant, regardless of the pathology, tends to have myoclonic seizures.

The 5 to 7 year old may start having absence attacks. The teenager tends to have generalized convulsions, and as the patient gets older, psychomotor attacks originating in the temporal lobe become more frequent. Seizures are more common in children than in adults. As children get older, the threshold for seizures increases and seizures tend to disappear.

<table>
<thead>
<tr>
<th>CAUSES OF EPILEPSY</th>
</tr>
</thead>
<tbody>
<tr>
<td>PREVENTABLE OR TREATABLE</td>
</tr>
<tr>
<td>Birth trauma and asphyxia</td>
</tr>
<tr>
<td>Infectious disease</td>
</tr>
<tr>
<td>Seizures following febrile convulsions</td>
</tr>
<tr>
<td>Toxic causes</td>
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<tr>
<td>Alcohol</td>
</tr>
<tr>
<td>Drugs</td>
</tr>
<tr>
<td>Pregnancy complications</td>
</tr>
<tr>
<td>Trauma</td>
</tr>
<tr>
<td>Cold injury</td>
</tr>
<tr>
<td>Metabolic and nutritional disorders</td>
</tr>
<tr>
<td>Endocrine disorders</td>
</tr>
</tbody>
</table>

Fig. 1

5 FACTORS TO CONSIDER IN SEEKING THE CAUSE OF EPILEPSY

1. NATURE AND SEVERITY OF BRAIN INSULT
2. AGE OF THE PATIENT
3. GENETIC BACKGROUND
4. THE ENVIRONMENT (PHYSICAL AND EMOTIONAL)
5. CHANGES IN THE METABOLIC STATE

Fig. 2
There have been many attempts to classify the different types of seizures. Most of the classifications agree in indicating a group of partial (or localized) seizures & another group of generalized seizures. This helped scientists to understand the epileptic process and eventually led to better treatment. A simple way of dividing different types of seizures is as follows (Fig. 3):

<table>
<thead>
<tr>
<th>CLASSIFICATION OF SEIZURES</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>1. PARTIAL SEIZURES</strong></td>
</tr>
<tr>
<td>Motor seizures (jacksonian)</td>
</tr>
<tr>
<td>Sensory seizures</td>
</tr>
<tr>
<td>Special sensory (visual, auditory) seizures</td>
</tr>
<tr>
<td>Complex (psychomotor, limbic) seizures</td>
</tr>
<tr>
<td><strong>2. GENERALIZED SEIZURES</strong></td>
</tr>
<tr>
<td>Tonic-clonic or major motor (Grand Mal)</td>
</tr>
<tr>
<td>Absence seizures (Petit Mal)</td>
</tr>
<tr>
<td>Seizures unique to childhood: infantile spasm, febrile convulsions, atonic seizures (drop attacks)</td>
</tr>
<tr>
<td><strong>3. UNCLASSIFIED</strong></td>
</tr>
</tbody>
</table>

Even this may seem too complicated and one may want to simply say seizures are major or minor.

When one has not seen the seizure, perhaps this is the best one can do, but there is merit in trying, with a careful history from the patient or relatives, to differentiate the various types of attacks.

The treatment of recurring generalized convulsions is different from absence attacks. A febrile convolution associated with an infectious disease in a child may never recur and treatment is not necessary. A pseudo-seizure is not a true epileptic attack; a teenager having such attacks is better treated with understanding and psychiatric help than with large amounts of anticonvulsants.

**THE AURA**

The aura is considered to be the onset of the attack, and is the first warning to the patient that an attack is beginning. The type of aura depends, of course, on where the attack originates in the brain.

**PARTIAL (FOCAL) SEIZURES**

Any of several types of focal or partial seizures may remain localized in one region of the brain, or they may spread to other regions of the brain and develop into generalized
convulsions. Focal seizures are more common in adults than in children, and one must always consider the possibility that they are caused by a tumor, or in endemic areas, by parasitic infestation, notably T. sodium neurocysticercosis.

**MOTOR SEIZURES**

A motor seizure originates in one part of the motor cortex and may remain focal or spread to involve the rest of the motor cortex (jacksonian seizure). The attack starts with tonic or clonic movements of part of the arm or leg, usually peripherally, and spreads to involve the rest of the body (Fig. 4).

There are other types of motor seizures such as the adversive seizure: here there is turning of the eyes, head and body away from the side of the brain that is firing.

**SENSORY ATTACKS**

Sensory attacks involve the part of the brain where sensations of various types are appreciated. The attack may be a numbness of the face, arm or leg. It may be visual such as flashing lights, auditory such as familiar sounds or tune. Abdominal auras are a feeling in the stomach which may rise up to the chest and neck and be followed by a generalized convulsion.

**CONVULSION**

A major convulsion - the generalized tonic-clonic convolution usually starts with the tonic phase (Fig. 5).

**TONIC PHASE:** The patient may cry out, fall to the ground unconscious, and develop generalized tonic stiffness of the body. The head is retracted, the arms flexed, and the
legs are extended. The muscles of the trunk are in spasm so that respiration ceases and the patient may become cyanotic.

After a varying period of time, the clonic phase begins.

CLONIC PHASE: it consists of alternate periods of spasms and relaxation of muscles so that clonic movements of the face, trunk and extremities develop. During the clonic phase, the patient may bite his tongue or become incontinent; breathing is labored or jerky and at times may appear to stop completely. Cyanosis may be profound.

In the normal course of events, the motor component of the seizure subsides and the patient may gradually regain consciousness. The patient may be confused, complain of fatigue or headache, experience difficulty speaking, or have some residual paresis. This often is followed by sleep.

Information on the postictal period is important. Weakness on one side of the face or arm, or a residual aphasia, for instance, may give a clue about the side of the origin of the attack. Data on the postictal period are also important in distinguishing a true seizure from a pseudo-seizure. The person who has had a hysterical attack may not have postictal headache or confusion, and may not sleep after the attack.

Another type of generalized seizure is the myoclonic seizure, a series of shocklike contractions of a group of muscles or a “spasm” of the whole body.

INFANTILE MASSIVE SPASMS, a generalized form of epilepsy, are myoclonic seizures that are dependent on the degree of maturation of the brainstem (midbrain, pons and medulla). They occur in infants, disappear as the brain matures, and can be caused by a great variety of pathological conditions. The spasms are the response of the brain and spinal cord, at a certain stage of maturation, to diffuse brain damage or dysfunction.
In the newborn child, epilepsy manifests itself by localized jerks. With increasing age, infantile massive spasm and Salaam or drop fits appear, indicating a higher level of origin in the central nervous system.

One kind of massive spasm consists of a sudden spasm of the body, extension of the legs, flexion of the head and neck, and upward extension of the arms.

The atonic attack, or drop fit, is expressed by a sudden loss of tone: the child is sudden thrown (or falls) to the ground. Since children frequently injure their heads this way, protection with some type of helmet may be required.

Seizures may be also classified by age groups (Fig. 6).

**ABSENCE ATTACKS (petit mal)**

The absence attack is a specific type of minor seizure usually occurring in children between the ages of 5 and 15. It used to be called “petit mal”. There are simple absence attacks, and absence seizures with motor manifestation. The absence attacks begin suddenly with a change in, or loss of, awareness or responsiveness. There also may be what is clinically termed a blank stare, brief upward rotation of the eyes, and interruption of activity. There is no motor accompaniment or gross convulsive movement, and the patient's posture does not change. The attack ends quickly.

It is usually brief, but may vary in duration. Parents describe simple absence attacks well when they say that for a few seconds the child is “dans la lune” or daydreaming. Often they think nothing of these attacks until someone, like a teacher, brings the problem to their attention. The child may present having had a generalized tonic-clonic seizure, and
when the parents are questioned, may be found to have had absence attacks for months or years. Absence attacks may have minor motor accompaniment such as blinking of the eyes. The attacks are usually of short duration, but occasionally as the children get older, the attacks go on for a prolonged period and have been called absence or petit mal status.

For the most part, the patient is unconscious or unaware of surroundings during an absence attack. An electroencephalogram (EEG) during an attack reveals 3 per second spike and wave activity which is characteristic of the disorder. The patient can have short burst of spike and wave activity on the EEG for which no clinical seizure can be seen.

One characteristic feature of absence attacks is the ease with which attacks may be precipitated by hyperventilation. If the child is asked to breathe deeply for a minute or two, attacks may be seen: this test can be done in front of the parents to demonstrate a seizure.

**FEBRILE CONVULSION**

Febrile convulsions are not a specific type of convulsion, but they should be mentioned separately. They are usually generalized tonic-clonic convulsions in young children under 3 years, associated with fever. They only occur with fever and may be an isolated event, never to recur with further febrile episodes. Some of these children go on to have seizures without fever, and then are considered to suffer from epilepsy.

The difficult decision to make is whether to treat them or not. Brain damage can occur from the anoxia of a prolonged seizure associated with a high fever. For this reason one tries to prevent recurring episodes of high fever. If it is possible to do an electroencephalogram (EEG) some time after the attack, and if abnormal, then it is wise to start prophylactic anticonvulsant medication.

**PSEUDO-SEIZURES**

Pseudo-seizures are not true seizures, but an attempt by the patient to gain attention, or avoid some undesirable situation. The attacks usually occur in front of an audience, they tend to be prolonged, there is rarely incontinence, and the patient usually does not suffer injury. There is often purposeless thrashing around, which sometimes can be interrupted by a strong stimulus. The attack is rarely followed by headache, confusion, or the usual postictal phenomena. The EEG is usually normal. They are sometimes difficult to distinguish from frontal seizures (i.e., originating in the frontal lobes).
OTHER PAROXYSMAL DISORDERS

There are other episodic disorders of the nervous system that may be confused with epilepsy (Fig. 7).

<table>
<thead>
<tr>
<th>OTHER PAROXYSMAL DISORDERS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pseudo-seizures</td>
</tr>
<tr>
<td>Migraine</td>
</tr>
<tr>
<td>Syncope</td>
</tr>
<tr>
<td>Narcolepsy</td>
</tr>
<tr>
<td>Vertigo and dizziness</td>
</tr>
<tr>
<td>Breath-holding spells</td>
</tr>
</tbody>
</table>

Fig. 7

THE ETIOLOGY OF EPILEPSY

There are many causes of epilepsy. The seizure itself is just a symptom of an underlying disorder of the brain. Sometimes the cause is obvious such as the child with facial hemangioma (Sturge-Weber syndrome) or tuberous sclerosis with adenoma sebaceum of the face. In many instances no specific cause can be found in the history or physical examination. Causes can be classified in a number of ways. One is by age. Some of the more common causes of epilepsy, arranged by age, are shown in Fig. 8.

<table>
<thead>
<tr>
<th>NEWBORN</th>
<th>Birth trauma, cerebral anoxia, congenital abnormalities, metabolic abnormalities</th>
</tr>
</thead>
<tbody>
<tr>
<td>INFANT</td>
<td>Birth trauma, Infection, fever, genetic cause</td>
</tr>
<tr>
<td>PRESCHOOL CHILD</td>
<td>Genetic cause, fever, infection, trauma, poisoning</td>
</tr>
<tr>
<td>SCHOOL-AGE CHILD</td>
<td>Genetic abnormalities, infection, trauma, native medicines</td>
</tr>
<tr>
<td>YOUNG ADULT (10 to 20)</td>
<td>Genetic abnormalities, infection, trauma, metabolic abnormalities, tumor</td>
</tr>
<tr>
<td>ADULT (20 to 50)</td>
<td>Alcohol, tumor, trauma, stroke, parasitic infestation</td>
</tr>
<tr>
<td>OLD ADULT (50 plus)</td>
<td>Tumor, trauma, stroke, alcohol</td>
</tr>
</tbody>
</table>

Fig. 8
The causes of epilepsy are multiple. There may be one, or a combination of causes. A child with a birth injury may have attacks only when there is a fever. A man who has had a head injury may have attacks only after drinking. A child with a strong family history of epilepsy may have seizures only when looking at the sun reflecting off water. One must look both for primary causes of brain injury or pathology and for secondary precipitating factors.

Many of the causes of brain damage that lead to epilepsy - birth injuries, severe convulsions with high fever, infections and head trauma - can be prevented. In epilepsy, as in other illness, it is better to prevent than to treat.

INVESTIGATION

A patient presents having had a convulsion. How does one proceed to investigate such a problem, so that one will have sufficient facts to treat the patient intelligently? (Fig. 9).

<table>
<thead>
<tr>
<th>PATIENT EVALUATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>HISTORY</td>
</tr>
<tr>
<td>PHYSICAL EXAMINATION</td>
</tr>
<tr>
<td>LABORATORY TESTS</td>
</tr>
<tr>
<td>RADIOLOGICAL STUDIES</td>
</tr>
<tr>
<td>ELECTROENCEPHALOGRAPHY</td>
</tr>
<tr>
<td>PSYCHOLOGICAL EVALUATION</td>
</tr>
<tr>
<td>SOCIAL BACKGROUND</td>
</tr>
</tbody>
</table>

Fig. 9
**History**

The most important first step is a careful history. This involves a description of the seizure, the frequency, precipitating factors, concurrent illnesses, etc. The familial history, and the past medical history which includes an account of the birth and early development, childhood diseases, school record, operations, a functional inquiry by systems, and social habits is elicited. By this time the recorder has made a rough estimate of the patient's intellectual ability.

**Physical examination**

The physical examination consist of a search for clues that might indicate a possible cause for a cerebral abnormality. Physical signs may be found in a patient with focal epilepsy. These signs may lead to a diagnosis of microcephaly, tuberous sclerosis, infantile hemiplegia, Sturge-Weber syndrome, or other neurological disorders. Subcutaneous nodules may point towards T. sodium neurocysticerciosis.

**Laboratory examination**

The majority of patients with epilepsy can be treated solely on the basis of findings in the history and physical examination. However, when possible, further studies should be done to clarify the type of epilepsy and to determine the basic cause that may require special treatment other than anticonvulsant. For example, if the seizures are due to tuberculosis of the brain, the patient must receive therapy appropriate for the tuberculosis as well as for convulsions. Whenever possible, the initial laboratory examination should include (Fig. 10):

- **Urine analysis**
- **Hemogram**
- **Blood chemistry**
- **Stool**
- **Mantoux test**
- **Radiological investigation**

*Fig. 10*
If possible, X rays of the skull should be done. In older patients, X rays of the chest are added, looking for tumor or tuberculosis. There are other radiological examinations such as computerized tomographic (CT) scan and magnetic resonance imaging (MRI) but usually are not available, and one must treat the patient without them.

Electroencephalography (EEG)

In the study of epilepsy, particularly in children and patients from whom one cannot obtain a good history, electroencephalography is very helpful in establishing the clinical diagnosis.

Psychological evaluation

The examiner always makes his own evaluation of the patient's intelligence. A more accurate study by a psychologist may be of value in determining whether the patient has an organic brain disorder. A patient of lower intelligence is less likely to take medication regularly, and this must be considered when planning therapy.

Social background

To complete the investigation of a patient with epilepsy, an understanding of the patient's social background is essential. Where does the patient live? What are the parents like? What type of assistance is there and what is the attitude of those at home? Knowledge of the background enables the physician to help the patient accept the handicap of epilepsy, to take medication regularly, and to adjust in society.

SUMMARY

When making a diagnosis in a patient with epilepsy, one attempts to document the type of seizure, determine the cause, and evaluate the intelligence and social background. This information, in turn, leads to correct treatment.

TREATMENT

There are many different aspects to treatment. The first is the handling of a patient having a generalized convulsion.

– How does one protect the patient from injury during the tonic-clonic unconscious phase?
- How does one prevent brain damage during status epilepticus?
- How does one stop status epilepticus?
- How are other types of seizures handled?

The attacks being over, medication must be prescribed to prevent recurring attacks in the immediate and distant future.

**GENERALIZED CONVULSION**

During the seizure the patient may fall, stiffen, and make jerking movements. A pale or bluish appearance may result from difficult breathing. Immediate care should be aimed at keeping the patient comfortable and safe while the seizure runs its course (Fig. 11).

Specifically:

<table>
<thead>
<tr>
<th>Help the person into a lying position and put something soft under the head</th>
</tr>
</thead>
<tbody>
<tr>
<td>Remove glasses and loosen any tight clothing</td>
</tr>
<tr>
<td>Clear the area of objects that might harm the patient</td>
</tr>
<tr>
<td>Do not force anything into the patient’s mouth</td>
</tr>
<tr>
<td>Keep the patient on is or her side, if possible, so that mucus and saliva can run freely and food or vomitus can be expelled</td>
</tr>
<tr>
<td>Hold the chin up and keep the airway clean</td>
</tr>
<tr>
<td>Do not try to restrain the patient. You cannot stop the seizure.</td>
</tr>
</tbody>
</table>

*Fig. 11*
After the seizure, keep the patient on one side to allow the saliva to drain from the mouth. Do not offer food or drink until the patient is fully awake. If the patient has one seizure after another (status epilepticus), intravenous therapy may be necessary. If this is not feasible, intramuscular or rectal medication may control the attacks.

**STATUS EPILEPTICUS**

First, make sure that the patient is having genuine seizures.

Therapy for status epilepticus should be undertaken in 4 stages (Fig. 12):

1. **Supportive measures to include making sure there is a clear airway.**

2. **Stopping seizures with a short-acting agent such as diazepam.** This is given as an intravenous bolus of 0.2 to 0.4 mg/kg of body weight at a rate of 1 mg/min. to a maximum of 10 mg in a single injection.
   
   Do not mix or dilute. It may be repeated in 15 minutes and again 15 minutes later. Respiration and blood pressure should be watched. If diazepam is not available, a barbiturate or phenytoin may be used.

   Intravenous therapy is not always feasible. Diazepam solution (the same as given parenterally) may be given rectally. Give 0.5 mg/Kg. Draw up the diazepam solution in a tuberulin syringe, remove the needle, insert the syringe into the rectum and give the solution as a bolus. This may be repeated if the seizure activity lasts more than 10 minutes.

   If diazepam is not available, rectal paraldehyde 8 cc in adult or chloral hydrate may be given by rectum.

3. **Following this, and as soon as it is possible, loading doses of phenytoin, phenobarbital, primidone or carbamazepine should be started.** The choice depends on what is available, the age and size of the patient, etc. This is followed by maintenance therapy.

4. **After emergency treatment of status epilepticus, the underlying cause should be searched for and treated.** Precipitating conditions include meningitis, encephalitis, head trauma, cerebral infarction, brain tumors, alcohol or drug withdrawal or metabolic disturbances.

   Remember that status epilepticus may also be precipitated by sudden withdrawal of anticonvulsant medication.
STATUS EPILEPTICUS: PHASES OF THE THERAPY

1. GIVE SUPPORTIVE MEASURES
2. STOP THE SEIZURE (EMERGENCY THERAPY)
3. START WITH MEDICATION (MAINTENANCE THERAPY)
4. LOOK FOR THE CAUSES OF THE STATUS

Fig. 12

PSYCHOMOTOR SEIZURES

During the seizure the patient may have a glassy stare, give no response or an inappropriate response when questioned, sit, stand, or walk around aimlessly, make lip smacking or chewing motions, play with clothes, or appear to be drunk, drugged or even psychotic.

In such cases:
- do not try to restrain the patient
- remove harmful objects or coax patient away from them
- do not agitate the patient
- when alone, be cautious when approaching the patient, particularly if he or she appears angry or aggressive.

After the seizure the person may be confused or disoriented and should not be left alone until fully alert. When the patient is fully awake, it is usually wise to give an extra dose of his or her regular medication. Very often the seizure has resulted from the patient having forgotten to take the medication.

LONG-TERM THERAPY

Anticonvulsant compounds are the backbone of therapy. The aim of treatment is to prevent seizures without producing toxic side effects.

Select the drug most likely to help your patient.

Certain compounds such as ethosuximide and valproic acid are more effective in preventing absence attacks.
Generalized tonic-clonic seizures and psychomotor seizures respond better to phenytoin, carbamazepine, phenobarbital, primidone, valproate, etc.

The selection of the anticonvulsant compound may depend on what is available on a continuous basis and the ability of the patient to pay. This is why phenobarbital is often the drug of choice.

Monotherapy, or use of one anticonvulsant alone, is the most effective and satisfactory form of therapy.

Compliance: there is no use prescribing medication if the patient does not take it regularly. The patient must be given instructions in writing, and the importance of taking it regularly must be stressed. It may help to divide the weeks supply of medicine into daily lots, to be sure that each day the prescribed amount is taken.

The ability to measure serum levels of anticonvulsants has shown that many patients do not take their medication. Look for and treat the primary underlying cause.

Why medication fails: in spite of the physician's best efforts, some patients fail to respond to medication and continue to have seizures. There are several possible explanations and each one must be considered:

- The patient does not take the prescribed medication on a regular basis, either because he or she forgets, does not like it, or feels better when not taking it.
- The dosage is not large enough, so there is not enough of the drug in the blood and the serum levels are below the effective range.
- The patient or the parents cannot afford the medication.
- Medication is not available. This often happens in isolated communities or developing countries.
- The wrong medication is being prescribed.
- The lesion or disorder causing the seizures is so severe that nothing will completely control the attacks.
Commonly used anticonvulsants

The following compounds are those most commonly used in the treatment of epilepsy (Fig. 13):

<table>
<thead>
<tr>
<th>DRUG</th>
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<tbody>
<tr>
<td>CARBAMAZEPINE</td>
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<tr>
<td>DIAZEPAM</td>
</tr>
<tr>
<td>ETHOSUXIMIDE</td>
</tr>
<tr>
<td>NITRAZEPAM</td>
</tr>
<tr>
<td>PARALDEHYDE</td>
</tr>
<tr>
<td>PHENYTOIN</td>
</tr>
<tr>
<td>PHENOBARBITAL</td>
</tr>
<tr>
<td>PRIMIDONE</td>
</tr>
<tr>
<td>TRIMETHADIONE</td>
</tr>
<tr>
<td>VALPROIC ACID</td>
</tr>
</tbody>
</table>

**Fig. 13**

**ADMINISTRATION**

**In the newborn**

In the newborn, one can usually start with elixir phenobarbital 3 mg/Kg. This preparation is easy to administer, and can be given every 2 hours depending on the response. Since elixir phenobarbital is detoxified and excreted quickly, fairly large doses may be necessary. If intravenous dosage is needed to control seizures, diazepam is the medication of choice. If the attacks are not easily controlled, the child should be sent to a referral centre.

**In infants**

In infants, one starts with elixir phenobarbital (15 to 60 mg per day). It is effective and usually all that is needed.

If the elixir is not available, pills (15 mg) can be crushed and given on a spoon. Medication can be given twice daily. Phenytoin is available as 50 mg flavored tablets which can be divided and crushed. Whether phenobarbital or phenytoin is chosen, the physician starts with a small daily dose and gradually increases the medication until the attacks are controlled, or there is evidence of toxicity. Before the child starts to walk, ataxia may
be difficult to recognize; accordingly, the child should be watched very carefully for signs of toxicity.

Other compounds are used for infants with special problems. For example, in children with infantile massive spasms, the physician starts with adrenocorticotopic hormone (ACTH) or prednisone, and if spasms are not controlled, add standard anticonvulsants, e.g., phenobarbital, phenytoin, primidone, nitrazepam, or carbamazepine.

**In children**

For focal and generalized tonic-clonic seizures, phenytoin infantabs 50 mg are most useful (100 or 200 mg per day). If this does not control the attacks, phenobarbital 30 mg can be added at bedtime. Medication can be given twice a day. The simpler the dosage and the fewer the doses per day, the better will be the compliance.

Other medications are available and may be most useful. Primidone may replace the phenobarbital. Valproic acid has proven to be useful in generalized epilepsy and should be tried in patients who are not controlled with other medications. Similarly, carbamazepine may be effective when used alone or in combination with other drugs. Both valproic acid and carbamazepine are metabolized quickly and it is better to give them three or four times a day rather than larger doses twice a day.

Absence seizures present a special problem. The response to ethosuximide may be very dramatic. This drug is available in capsules (250 mg) and sometimes as a syrup. It is wise to start slowly, increasing the dosage from 250 mg per day to 750 or 1000 mg per day. If the appetite is affected, the dose should be reduced. If the absence attacks are not controlled with ethosuximide, valproic acid should be tried. If there are associated focal or generalized seizures, pheno-barbital, primidone, phenytoin, valproic acid or carbamazepine may be added.

**In adolescents**

Teenage children can be treated with adult dose. The physician should start with one compound (usually phenytoin) and increase the dose until the seizures are controlled or toxicity develops. Then a second compound may be added, making changes slowly and trying to give as little medication as possible compatible with complete control.

Sometimes, particularly in partial seizures with automatism, one starts with carbamazepine 200 mg twice a day and slowly increases until control is achieved. There is a gradual increasing of the dose until adequate serum levels and control of seizures are obtained.

Valproic acid may also be successful in adolescent patients with generalized tonic-clonic or absence seizures.
Ethosuximide is still effective at this age for absence seizures, and can be given in combination with other drugs. Adolescent children often forget to take the medication or rebel and refuse to take it. This is often the cause of isolated seizures. One must try to determine whether the patient has been taking medication regularly, or has failed to comply. If poor compliance is the case, one should not increase the dosage, but rather should discuss the problem with the patient, stressing the importance of regular drug therapy.

When compliance is good and seizures still occur, one may want to increase the dosage, or try another compound. The ability to measure serum drug levels has been of tremendous help in determining whether a patient is taking medication or not.

**Toxic effects of anticonvulsants**

Toxic effects of anticonvulsants may be dose related or hypersensitivity reactions.

<table>
<thead>
<tr>
<th>DOSE-RELATED TOXICITY</th>
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<tbody>
<tr>
<td>DROWSINESS</td>
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<tr>
<td>NYSTAGMUS</td>
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<td>ATAXIA</td>
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<td>MENTAL CONFUSION</td>
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<tr>
<td>BEHAVIOR PROBLEMS</td>
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<tr>
<td>GUM HYPERTROPHY</td>
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</table>

Fig. 14

The secret of prevention is to start slowly and gradually increase the dosage until therapeutic levels are reached.

With phenobarbital the most frequent sign of overdose is somnolence and irritability; with phenytoin it is an unsteady gait; with carbamazepine, dizziness, double vision or headache; valproic acid often causes nausea, weight gain, or temporary loss of hair.
Hypersensitivity

Hypersensitivity causes skin rashes, urticaria and sore mouth.

The most serious reaction is the suppression of bone marrow activity and the development of agranulocytosis. Sodium valproate may cause liver toxicity. Performing regular blood studies may not be practical. The patient or relatives should be advised about possible toxic effects and advised to report immediately should these occur.

The treatment of epilepsy does not end with the prescription of anticonvulsant medication. The medical practitioner must advise and help the patient and family with the problems that arise. It may be schooling, when to drive a car, plans for employment, marriage, and many others.

The practitioner who develops a good rapport with the patient can be of great assistance, and should be prepared to spend time to listen, help, advise, educate. The reward is considerable.

- Epilepsy is a treatable disorder.
- Patients with epilepsy can be helped.
- The person with seizures is a person who should take his rightful place in society.
- Many great men and women had epilepsy and conquered it. Your patient can too.
- Epilepsy is primarily a disorder of children. With proper treatment they live to grow out of it.
- A medical person treating seizures has a major responsibility - but the reward is correspondingly great.
Chapter 7

PRIMARY PREVENTIVE NEUROLOGY

The potential of the Nervous System to recover lost function after an injury is poor when compared with many other organ systems. For this reason there is the need to emphasize the protection of the Nervous System from injury through disease by prevention at the primary health level.

Nutritional factors, infections, and toxic substances all affect the health of the Nervous System as they affect health generally (Fig 1).

1. NUTRITIONAL FACTORS

Preventive Neurology begins during pregnancy.

- Prematurity, with very low birth weight infants (500 to 1500 gms), results in cerebral palsy among 12% of survivors, but other major neurological handicaps (mental retardation, epilepsy, deafness, or blindness) increase this figure to 18%. Malnutrition, alcohol and cigarette smoking during pregnancy are other major causes of prematurity and need correction if babies are to be born with a fair chance to develop normally, free of major neurological disorders.
During infancy and childhood, good nutrition is essential for the final optimal development of the Nervous System, especially in reference to sufficient protein, vitamins and minerals intake. High quality protein, as in mother's milk or cow's milk, is required during the first 2 years while the Central Nervous System is developing rapidly.

Deficiency of certain minerals results in a serious neurological syndrome. Studies show that deficiency of iodine is a significant cause of endemic cretinism, a syndrome which is characterized by mental deficiency, deaf mutism, spastic diplegia and lesser degrees of neurological defects. It is estimated that between 0.6 and 1 billion people are at risk for iodine deficiency in various regions of the world, most of them in the developing countries, with 710 million in Asia, 227 in Africa, 60 million in Latin America and 20-30 million in Europe. Iodized salt and iodized oil (by injection or by mouth) are suitable for correction of iodine deficiency on a mass scale.

Vitamin deficiencies, either specific or in combination and together with other nutritional factors, can cause major neurological disturbances.

Deficiency of vitamin A has high prevalence in Asia especially among children, leading to several visual problems. Primary prevention is dependent on a diet that contains adequate vitamin A. Dairy products containing butter, egg yolk, liver, and fish liver oils are excellent sources. In tropical and developing countries, fruits, such as mango and papaya, and African palm oil used in cooking are rich in vitamin A.
VITAMIN A
RECOMMENDED INTAKE

<table>
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<tr>
<th>ADULT MALES</th>
<th>1000 µg RETINOL per day</th>
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<tr>
<td>ADULT FEMALES</td>
<td>- 800 µg RETINOL per day</td>
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<tr>
<td></td>
<td>- 1000 µg during pregnancy</td>
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<td>- 1200 µg during lactation</td>
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**Fig. 4**

The specific deficiency of vitamin B1, thiamine, is the main cause of the Beriberi and Wernicke-Korsakoff syndrome.

Beriberi occurs primarily where polished rice is the main staple food or alcohol is consumed in large amounts. It is characterized by progressive weakness and muscle wasting involving symmetrically the legs more than the arms and distal muscles more than the proximal ones. Weakness may become so severe as to result in paralysis of legs and incapacitated hands (see peripheral neuropathy). Abnormalities of sensation are usually prominent and complaints of aching, coldness, hotness, numbness, or paresthesiae with tenderness of the calves, soles of feet and fingers. Pressure or even light touch may become painful as muscles become wasted and atrophic. Thus, severe nutritional polyneuropathy with both sensory and motor disturbances is present.

The Wernicke-Korsakoff syndrome is also a manifestation of thiamine deficiency and of particular importance in severe alcoholics, most of whose calories are derived from alcohol. With the Wernicke-Korsakoff syndrome, disturbed ocular motility with nystagmus, ataxia and impaired mentation are prominent. Memory is particularly affected in the Korsakoff psychosis rendering some patients incapable of any but the simplest tasks.

**THIAMINE
RECOMMENDED INTAKE**

0.5 mg or more per 1000 calories per day

**Fig. 5**
Prevention of these two syndromes is possible by increasing the intake of thiamine. Enrichment of polished rice at the mill with thiamine, riboflavin and niacin would prevent much of the vitamin B diseases in the world and needs to be adopted in countries where polished rice is the major staple food. Alcoholics could be protected from deficiency states by enrichment of alcoholic beverages by these vitamins. Barring this, ingestion of whole grain, peanuts, pork, wheat germ or multivitamin supplements are preventive.

Niacin and tryptophan deficiency causes pellagra with glossitis, dermatitis of exposed skin and dementia. The demented person may be hyperactive and manic or apathetic, lethargic and obtunded. This deficiency occurs largely with corn-dependent diets which lack both niacin and tryptophan. Supplementing maize diets with sufficient legumen protein prevents the deficiency. The best food sources of niacin are yeast, liver, lean meat, poultry, peanuts, and vegetables.

**NIACIN RECOMMENDED INTAKE**

<table>
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<tr>
<th>6.6 NIACIN EQUIVALENTS per 1000 calories per day</th>
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<tr>
<td>(1 niacin equivalent = 1 mg of niacin or 60 mg tryptophan)</td>
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*Fig. 6*

**SOURCES OF NIACIN**

- Liver
- Peanuts
- Poultry
- Legumes

*Fig. 7*
2. INFECTIOUS DISEASES

A. Meningitis

During infancy and early childhood, infectious diseases are major hazards to the nervous system. Bacterial meningitis affects children most commonly under the age of 4.

The most common causative agents are streptococcus pneumoniae, haemophilus influenzae and meningococcus. Early treatment of middle ear infections and lung infections are necessary to prevent meningitis by other organism.

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<tr>
<th>ETIOLOGY OF ACUTE BACTERIAL MENINGITIS BY AGE</th>
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<tbody>
<tr>
<td><strong>NEONATAL</strong></td>
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<tr>
<td>Gram negative enterobacilli 60%</td>
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<tr>
<td>Streptococci group B 20</td>
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<tr>
<td>Staphylococcus aureus 5</td>
</tr>
<tr>
<td>Listeria monocytogenes 15</td>
</tr>
<tr>
<td><strong>INFANTS AND CHILDREN</strong></td>
</tr>
<tr>
<td>H. influenzae 40 %</td>
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<tr>
<td>S. Pneumoniae 20</td>
</tr>
<tr>
<td>N. meningitidis 40</td>
</tr>
<tr>
<td><strong>ADULTS</strong></td>
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<tr>
<td>S. Pneumoniae 45 %</td>
</tr>
<tr>
<td>S. aureus 15</td>
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<tr>
<td>N. meningitidis 25</td>
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<tr>
<td>Streptococci 10</td>
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<tr>
<td>Gram negativebacilli 5</td>
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</table>

**Fig. 8**

B. Measles

Measles are highly contagious viral disease which kills several hundred thousand infants and young children yearly. Essentially all children will become infected and at least 1 million children living in developing countries will die unless protected by immunization. Generally the peak incidence is under the age of 3 years. The youngest and most undernourished children suffer the most severe complications. Diarrhea, malnutrition, pneumonia and blindness associated with vitamin A deficiency are among the worst complications. Subacute sclerosing panencephalitis will be a sequelae in a few. The infection is preventable by the timely administration of a vaccine.
C. Poliomyelitis

With the development of successful immunization for poliomyelitis, this crippling infectious, viral disease affecting children has almost been eradicated in countries where mass immunization programs have been undertaken with trivalent vaccines. The virus infects and kills the anterior horn motor neurons in the spinal cord, leaving denervated muscles to atrophy (Fig. 10).

The paralysis can be extensive or selectively affect only a small number of muscles. Bulbar polio, including muscles of respiration, has been the most feared form of the disease. The major obstacle to control of poliomyelitis is failure to immunize susceptible children.
D. Leprosy

Leprosy is still a prevalent disease with around 2 to 3 million people being disabled by it primarily in tropical and subtropical developing countries. Many developing countries consider leprosy a major health problem because a significant proportion of cases result in deformity and subsequent social stigmatization. The deformities result from the predilection of the infecting bacteria, mycobacterium leprae, for sensory nerves. With loss of sensation, repetitive unrecognized trauma results in tissue destruction and deformities. Transmission of infection is by direct and prolonged contact. Prior to availability of chemotherapy, isolation in colonies was the only known way to prevent spread of the disease. Generally, household contacts of patients with lepromatous or borderline lepromatous leprosy - forms of the disease in which the causative bacilli are numerous in the skin lesions - should be considered for dapsone prophylaxis.

3. TOXIC SUBSTANCES

There is a large and increasing number of chemical toxins which adversely affect the nervous system.

Many medications and drugs are potential neurotoxins. Health professionals must be aware of such toxicity and monitor their patients accordingly.

A. Lead neuropathy

Environmental contamination with lead has increased greatly in industrialized countries. Contamination of lead may be a result of different sources. Many occupations entail significant exposure. Smelter workers, miners, storage battery workers and pottery makers are particularly heavily exposed. Workers in automobile manufacturing, ship building, paint manufacture, and the printing industry may also experience toxic exposure.

Tetraethyl lead in gasoline causes poisoning through inhalation and this organic compound seems to have a proclivity for the central nervous system.

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<th>INDUSTRIES USING FOR PRODUCING LEAS</th>
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<tr>
<td>Petroleum, mining and smating,</td>
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<tr>
<td>storage battery manufacture,</td>
</tr>
<tr>
<td>printing, paint and pigment,</td>
</tr>
<tr>
<td>ceramic and glass, construction,</td>
</tr>
<tr>
<td>ammunition, wrecking and salvage,</td>
</tr>
<tr>
<td>battery reclaiming, brass poishing.</td>
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</table>

Fig. 11
Either the brain or peripheral nerves may be affected by chronic lead poisoning. Encephalopathy with papilledema and increased intracranial pressure are late stages of CNS involvement.

Early symptoms may be vague and include irritability, incoordination, memory lapse, labile affect, paranoia, headache, lethargy and dizziness.

In more severe poisoning syncope, disorientation, flaccidity, mental impairment, ataxia, psychosis, blindness and coma may occur.

Peripheral nerve involvement is seen more commonly in adults than in children, and is exclusively motor. Wrist drop and foot drop which may be asymmetrical are seen most often.

Prevention is dependent upon recognition of potential sources of poisoning and avoiding exposure to such sources. Awareness of associated intestinal colic and hemolytic anemia may allow early recognition and prevention of neurological complications. Early treatment with appropriate chelating agents such as BAL (dimercaprol), versine or D-penicillamine, may prevent later neurological disease.

B. Mercury Intoxication

Methyl mercury is readily absorbed from the intestinal tract and distributes widely in the body. It passes through the placenta into the fetus and about 10% localizes in the brain.

The resulting brain damage is largely irreversible. Major epidemics have followed industrial contamination of water with subsequent biotransformation of inorganic mercury to methyl mercury to be ingested by fish and man.

Grass contaminated by organic mercurial pesticides has also caused intoxication. Such intoxication is associated with high death rates and permanent brain damage in survivors. Neurological symptoms include severe paresthesiae, dysrhythmia, ataxia, visual field constriction, deafness, blindness, spastic paralysis, dementia and coma. Children of methyl-mercury poisoned mothers may show various degrees of cerebral palsy-like abnormalities and mental retardation.

C. Oxygen poisoning of the newborn

In the newborn, where administration of oxygen is imperative, administration of high oxygen concentrations can cause retrolental fibrosysplasia and blindness. Arterial hyperoxia causes severe vasoconstriction of retinal vessels with ischemic necrosis and fibrosis of the retina. This vasospastic response to high arterial oxygen tension is restricted to retinal vessels and only to the neonatal period.
4. **OTHER DISEASES with neurological implications are recognized as a preventable major health problem.**

A. Convulsions

Epilepsy has many causes some of which can be prevented. For example: birth trauma, head trauma from motor vehicle accidents, infections, such as meningitis or brain abscess, cysticercosis (the larvae of pork tape-worm) and cerebral vascular accidents. It may result also from severe alcohol or other drug abuse and withdrawal.

Convulsions also often occur in small children as a result simply of high fever without any underlying brain pathology. This type of convulsions may be followed, at a later time, by a seizure disorder.

B. Cerebro-Vascular Accidents

Strokes or cerebro-vascular accidents (CVA) are a major cause of death and disability throughout the world. Developed and developing countries are all affected. Strokes result from blockage of blood vessels by clots or atheroma or from rupture of vessels with hemorrhage. A major cause of such vascular accidents is systemic arterial hypertension, that is: high blood pressure.

High blood pressure most commonly begins in middle age, 40 years or older, when arterial pressure, as measured most commonly in the arm with an inflatable occluding cuff, rises above 90 mm Hg diastolic and/or above 140 mm Hg systolic pressure. Blood pressure above these normal limits increases the risk of strokes and heart attacks.

Although there are a few systemic diseases of the kidneys or endocrine system which may cause hypertension, the vast majority of cases are of essential hypertension, in whom genetic factors play a role, and overconsumption of sodium, over prolonged periods of time, contributes.

Strict salt restriction will reduce blood pressure in most hypertensive subjects. Much can be done to prevent strokes. In the past two decades there has been a 50% decrease in death rate from stroke in the U.S.A. secondary to a decrease in hypertension in the population. Education to increase the awareness of the prevalence and risks of hypertension, together with mass screening programs to identify individuals who have high blood pressure, have been undertaken.

For some persons with high blood pressure regular medication to reduce pressure is lifesaving. There are fortunately several effective pharmacologic approaches to management which physicians can prescribe. These require lifetime adherence and since some side effects are almost unavoidable with chronic medication use, every attempt should be made to reduce blood pressure by non-drug means. Reduced sodium in the diet, weight reduction, regular aerobic exercise, avoidance of heavy alcohol ingestion, and stress reduction can often normalize blood pressure in individuals with mild levels of hypertension. Fortunately, these simple measures can be effective but they do require major readjustments in long term behavior patterns in order that the reduction in pressure be sustained.
Chapter 8

GLOSSARY OF NEUROLOGICAL TERMS

A

ABSCESS (CEREBRAL) localized collection of pus within the brain parenchyma

AGNOSIA inability to recognize an object or to interpret a sensory stimulus even though the sensory system is preserved

AMAUROSIS loss of vision

AMNESIA loss of memory

ANEURYSM abnormal dilatation or expansion of the wall of an arterial vessel

APHASIA language disturbance

ARRHYTHMIA (CARDIAC) irregular heart beat

ASYMPTOMATIC producing no symptoms

ATAXIA loss of coordination

ATHEROSCLEROSIS vascular disorder with damage to the arterial walls

ATHETOSIS slow and irregular involuntary movements of the arms, legs and face

ATROPHY decrease of tissue mass
<table>
<thead>
<tr>
<th><strong>BABINSKI SIGN</strong></th>
<th>abnormal superficial reflex with dorsiflexion of the great toe after stimulation of the plantar surface of the foot</th>
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<tr>
<td><strong>BILATERAL</strong></td>
<td>on two sides</td>
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<tr>
<td><strong>BRADYKINESIA</strong></td>
<td>slowness of motor activities</td>
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<tr>
<td><strong>CEREBROSPINAL</strong></td>
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<tr>
<td><strong>CERVICAL</strong></td>
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<tr>
<td><strong>CHEYNE-STOKES RESPIRATION</strong></td>
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<td><strong>CHOREA</strong></td>
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<td><strong>COMA</strong></td>
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<td><strong>CONGENITAL</strong></td>
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<td><strong>CONTUSION</strong></td>
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<td>(MUSCULAR) DYSTROPHY</td>
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<td>ED</td>
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<td>EDEMA</td>
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<td>EMBOLISM</td>
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<td>EPILEPSY</td>
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