VOLUME 2

Pediatric Otiatrics

VOLUME
2

Pediatric Orthopedics

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Levels of Affection

The neuromuscular system may be affected at various levels, each of which is characterized by changes in motor function peculiar to the site and extent of involvement.

At the *spinomuscular level* motor activity is simple; the impulses arising in the anterior horn cells of the spinal cord are transmitted through the peripheral nerves to the myoneural junctions and then to the individual muscles. In disorders at the spinomuscular level, the loss of motor power is focal and segmental, with complete paralysis of the muscles or muscle groups that are supplied by a peripheral nerve or by the anterior horn cells in the spinal cord. Muscular paralysis is flaccid or hypotonic, with reaction of degeneration, atrophy, fibrillations, and fasciculations. The deep tendon and superficial reflexes are diminished or absent. Pyramidal tract signs, abnormal involuntary movements, and ataxia are absent. There may be trophic changes in the skin, nails, and bone.

Pathologic processes at the spinomuscular level may be further classified into various sublevels. When the disease originates in the anterior horn cells, as in poliomyelitis, the *spinal level* of the motor system is affected. Other examples of diseases at the spinal level are: progressive spinal muscular atrophy of the Werdnig-Hoffmann type, progressive bulbar palsy, syringomyelia, and intramedullary neoplasm. The loss of function of the anterior horn cells and the motor nuclei of the brain stem results in clinical findings of flaccid paralysis, atrophy, areflexia, reaction of degeneration, and fasciculations.

At the *neural level* of the motor system, the peripheral nerves and nerve roots are affected, common examples of which are obstetrical brachial plexus palsy and progressive neural muscular atrophy (Charcot-Marie-Tooth disease). In affections of nerves sensory fibers are usually involved, with resultant sensory changes such as anesthesia or hyperesthesia. Otherwise, the clinical findings are similar to those of spinal level affections, i.e., there is flaccid paralysis, atrophy, reaction of degeneration, and areflexia as a result of loss of conduction of motor impulses. In the absence of sensory changes it is difficult to distinguish between diseases of the peripheral nerves, anterior roots and anterior horn cells.

When the pathologic process arises at the myoneural junction, as in myasthenia gravis and familial periodic paralysis, then it is a disease at the *myoneural level.* In diseases of primarily muscular origin, the motor system is involved at the *muscular level.* The muscular dystrophies are familiar examples of disturbance of the muscular level in disease at the spinomuscular level. Paralysis is flaccid, but reflexes persist until the late stages, when marked atrophy has occurred. There is loss of contractibility without loss of excitability, i.e., the muscle fibers have degenerated and have been replaced by fibroadipose tissue, but the peripheral nerves and anterior horn cells are normal.

In disorders of the motor system at the *extrapyramidal level* there is generalized involvement of the muscles of the limbs and trunk. The muscle tone is hypertonic. Atrophy, fasciculations, and reaction of degeneration are absent. Motion of the limbs is hyperkinetic, with loss of associated or automatic movements. The deep tendon and superficial reflexes are normal. There are no pyramidal tract responses and no sensory deficit. Athetoid cerebral palsy is a common example of a disease at the extrapyramidal level.

At the *pyramidal* or *corticospinal level* of involvement, motor deficit arises from affection of motor nuclei of the cerebral cortex. Paresis is usually generalized and associated with hypertonicity of spasticity of muscles. Pyramidal tract signs and pathologic reflexes are usually present. There is usually some atrophy that is not focal; it is caused by chronic paralysis and disuse. Fasciculations, trophic disturbances, reaction of degeneration, and abnormal movements are absent. The deep tendon reflexes are hyperactive and the superficial reflexes are diminished or absent. Spastic cerebral palsy illustrates the pyramidal level of motor involvement.

Cerebellar level lesions are characterized by loss of coordination and control, or ataxia. There is no real loss of motor power. Fasciculations, reaction of degeneration, atrophy, or trophic disturbances are absent. The deep tendon reflexes may be diminished or pendular, but the superficial

reflexes are normal. Pyramidal tract responses cannot be elicited.

The *psychomotor level* of motor performance is the highest level of neuromuscular activity—at which volitional movements are initiated and affected by integration, memory, and symbolization. Paralysis caused by hysteria is an example of psychomotor disturbance. Loss of motor power is bizarre, with no actual paralysis. There is no real neurologic deficit. There are no fasciculations, no atrophy, and no true ataxia.

Differential features of various levels of motor function are illustrated in Table 5–1.

Neuromuscular System as a Functional Unit

Muscles are the expressive unit of the neuromuscular system and the moving force of the body. Muscles whose contraction directly produces a specific action are classified as *agonists* or *prime movers* (protagonists). An example is the biceps brachii in flexion of the elbow. Those muscles that oppose the agonists must be relaxed for contraction of the agonists (these are called *antagonists* or *moderators*) as, for example, the triceps brachii is in flexion of the elbow.

A motor action, even in an apparently simple motion, is quite complex. It involves the *muscles of fixation*, which stabilize the adjacent joints and afford a firm base for muscle action. The action of *synergists* is to assist the agonists and to reduce to a minimum all unnecessary motions. The execution of a motor movement requires the coordinated action of all four physiologic muscle groups—the contraction of agonists and the relaxation of antagonists as well as the associated function of the synergists and the muscles of fixation. Loss of function of any of these muscle groups will result in disturbance of motor performance.

Responses of Muscles

The responses of muscles to injury and disease are predictable. Muscles that are not used atrophy. The rapidity of development of such disuse atrophy is well illustrated by the atrophy of the quadriceps femoris that follows a painful lesion of the knee or immobilization of the knee in a long leg cast. With progressive resistive exercises, muscles hypertrophy. Painful stimuli will cause protective spasm of a muscle, which, when maintained in its shortened position for a period of time, will tend to develop myostatic contracture. The antagonist muscles to those in spasm are weakened by being maintained in their longer, stretched position and by inhibition of their function and recovery.

Muscular action affects bone growth. In the growing skeleton, muscle imbalance will cause deformity in the direction of action of the stronger muscle. Muscles are very sensitive to ischemia, as illustrated by Volkmann's ischemic contracture. Chronic systemic disease causes generalized muscle weakness and increased fatigability.

Affections of the Brain and Spinal Cord

CEREBRAL PALSY

Definition

Cerebral palsy is difficult to define, as it is not a single disease entity but, rather, a convenient category denoting conditions having certain common characteristics. The generally accepted criteria of the symptom complex of cerebral palsy are as follows:

1. It must be due to a fixed, nonprogressive brain lesion or lesions. There should not exist any active disease at the time of diagnosis. Thus, transient disorders or those that are the result of a progressive disease of the brain or spinal cord are excluded.

2. The original lesion must occur prenatally, at birth, or early in the postnatal period. The exact limits of this early period are not agreed upon, and it is best to avoid arbitrary age limits. The interference with the developing central nervous system by the early fixed lesion is the significant pathologic feature.

3. In certain children, the primary disorder involves the musculoskeletal system

Table 5–1. *Differentiation of Motor Disorders at Various Levels of Neuromuscular Function**

	Spinomuscular			Extrapyramidal	Pyramidal	Cerebellar	Psychomotor
	Muscular	*Neural*	*Spinal*				
Loss of motor power	Focal-segmental Usually proximal and axial muscle groups	Focal-segmental Usually distal limb musculature	Focal-segmental Usually distal limb musculature	Generalized Entire limb and movements	Generalized Entire limb and movements	None Ataxia may simulate loss of power	No true loss Bizarre, may simulate any type
	Complete	Complete	Complete	Incomplete	Incomplete		
Tone	Flaccid	Flaccid	Flaccid	Rigid	Spastic	Hypotonic (ataxia)	Normal or variable, may be increased
Atrophy	Present	Present	Present	Absent	Minimal (due to disuse and chronic paresis)	Absent	Absent
Fasciculations	May be present	Absent	May be present	Absent	Absent	Absent	Absent
Reaction of degeneration	Present	Present	Present	Absent	Absent	Absent	Absent
EMG							
Interference pattern	Normal until late in disease	Reduced	Reduced				
Fibrillation potential	Not usually present	Present	Usually present				
Action potential	Short duration	Prolonged with normal or polyphasic potentials	Prolonged with occasional giant potentials				
Evoked sensory and mixed nerve potentials	Normal	Absent, diminished amplitude, or prolonged conduction time	Normal				

Reflexes							
Deep	Diminished and preserved until late	Absent early	Absent early	Normal or variable	Hyperactive	Diminished or pendular	Normal or increased range
Superficial	Diminished	Absent	Absent	Normal or increased	Diminished or absent	Normal	Normal or increased
Pyramidal tract response	No	No	No	No	Yes	No	No
Sensory deficit	Absent	Usually present	Absent	Absent	May be present (stereognosis or other cortical)	Absent	Absent
Trophic disturbance	Present	Present	Present	Absent	Usually absent	Absent	Absent
Ataxia	Absent	Absent	Absent	Absent	Absent	Present	Absent (may simulate ataxia)
Abnormal movements	Absent	Absent	Absent	Present	None	May be present (intention tremor and ataxia)	May be present
Associated movements	Normal	Normal	Normal	Absence of normal associated movements	Presence of pathologic associated movements	Normal	Normal

*Adapted from DeJong, R. N.: The Neurological Examination. 3rd edition. New York, Hoeber Medical Division, Harper & Row, 1967, p. 382; and Farmer, T. W.: Pediatric Neurology. New York, Hoeber Medical Division, Harper & Row, 1964, p. 612.

and lack of motor control is the greater handicap, whereas in others mental retardation, convulsions, sensory disturbance, speech impediments, or defects of hearing, language, or eyesight may be the more important difficulty.

The term *cerebral palsy* has certain administrative usefulness. The foregoing criteria of this category of disease should, however, be carefully examined, as conditions such as Friedreich's ataxia, progressive hereditary paraplegia, or amaurotic familial idiocy should not be included under the heading of "cerebral palsy."

Classification

The differing approaches of clinicians and therapists concerned with the diagnosis and treatment of cerebral palsy are reflected in the various classifications they have used.[2, 5, 75, 83, 186] During the past 25 years, the ones most commonly employed are essentially modifications of Phelps's description of the clinical manifestations of cerebral palsy, which consists primarily of helpful suggestions to therapists and others concerned with the practical management of these patients.[142–144] Phelps based his classification primarily on the state of muscle tone, the presence or absence of involuntary movement, and the topographical distribution of motor deficits, taking into account etiologic factors, the presumed site of neuropathologic changes, and associated sensory defects (Table 5–2).

The defect of a classification that defines categories primarily in terms of changes in muscle tone is that the muscle tone of individual patients varies greatly with maturation and may alter considerably from day to day—and even from hour to hour—according to position, posture, state of alertness of fatigue, environmental temperature, and emotional state.

Perlstein and Minear have attempted to produce more comprehensive descriptive classifications by considering the site of pathologic change, clinical manifestations, topographical description, severity of motor involvement, muscle tone and etiology. (Tables 5–3 and 5–4).[127, 138, 139]

Crothers and Paine, stressing that the characteristic signs in cerebral palsy manifest themselves only gradually, have offered a more explicit neurologic classification (Table 5–5).[49]

Table 5–2. *Recent American Classification of Cases of Cerebral Palsy**

Spastic	
Aspastic	
Spastic	
Monoplegia	
Hemiplegia	
Paraplegia	
Triplegia	
Quadriplegia	
Basilar	
Athetosis	
Tension	Rotary
Nontension	Emotional release
Dystonic	Tremor
Flail	Unclassified
Arm neck	Paraplegia
Deaf	Quadriplegia
Shudder	Monoplegia
Hemiathetoid	Recovered
Cerebellar release	
Rigidity	
Intermittent	
Continuous	
Miscellaneous	
Hemiplegia	
Paraplegia	
Triplegia	
Quadriplegia	
Tremor	
Intention	
Constant	
Ataxia	
Cerebellar	
Eighth nerve	

*Based on that of Phelps by Hellebrandt, F. A.: Trends in the management of cerebral palsy. Lectures in Medical College of Virginia (unpublished manuscript). 1950–1951.

Ingram and Balf and Ingram have suggested a classification by neurologic syndromes based on that of Freud, with modifications made necessary by advances in knowledge since his time (Table 5–6).[12, 89]

Classification of cerebral palsy is difficult but very important. There is no general agreement. In the care of the child with cerebral palsy, the orthopedist is part of a multidisciplinary team. The neurologist or pediatrician may be using any one of the foregoing classifications, and the orthopedic surgeon should be familiar with their vocabulary.

The distribution of paralysis is described as follows, according to the number of

Table 5–3. *Classification of Perlstein, 1952**

By Clinical Symptoms	*Topographical Involvement of Extremities*	*By Muscle Tone*	*Severity*	*Etiology*
Spastic conditions	Paraplegia	Isotonic	Mild	Prenatal
				Hereditary
Dyskinesias	Diplegia	Hypertonic	Moderate	Static
Choreas	Quadriplegia or tetraplegia	Hypotonic	Severe	Progressive
		–	–	Acquired in utero
Athetoids	Hemiplegia	–	–	Infection
Dystonia	Triplegia	–	–	Anoxia
Tremors	Monoplegia	–	–	Cerebral hemorrhage
Rigidity	Double hemiplegia	–	–	Rh factor
Ataxia	Limited to both upper extremities	–	–	Metabolic disturbance
				Gonadal irradiation
				Natal factors
				Anoxia
				Cerebral hemorrhage
				Trauma
				Pressure change, etc.
				Postnatal factors
				Trauma
				Infections
				Toxic causes
				Vascular accident
				Anoxia
				Neoplasms and developmental defects

*From Perlstein, M. A., and Barnett, H. E.: Nature and recognition of cerebral palsy in infancy. J.A.M.A., *148*:1389, 1952.

limbs involved. If a patient has one limb involved, the condition is termed *monoplegia;* if two limbs on the same side are affected, *hemiplegia;* if two legs, *paraplegia;* if three limbs, *triplegia;* or four limbs, *quadriplegia* or *tetraplegia.* There is little agreement about the use of the words *diplegia* or *double hemiplegia.* The term "cerebral diplegia" or "diplegia" is employed by some authors to describe the condition of patients with more or less symmetrical paralysis, dating from birth or shortly afterward, which is more severe in the lower than in the upper limbs.[48, 64, 88] *Double* or *bilateral hemiplegia* is used when the arms are more severely affected than the legs or when there is asymmetry of involvement.

Etiology and Pathology

The nonprogressive brain lesion in cerebral palsy may be due to birth injury, developmental malformations, or damage acquired postnatally. Etiologic diagnosis is circumstantial in the majority of patients. Pathologic findings are available in only a few patients, and even then, one has difficulty in determining the primary underlying cause. Ingram and Crothers and Paine have given a critical etiologic analysis of their own cases, as well as a comprehensive review of the literature.[49, 89]

BIRTH INJURY

Little first described three types of paralysis that could occur as a result of abnormal birth, denoting them as "hemiplegic rigidity," "paraplegia," or "generalized rigidity," as well as a condition characterized by "disordered movement."[115–117] These would now be considered forms of cerebral palsy.

Birth injury is direct or indirect damage of pregnancy or during labor and the process of delivery. It must be differentiated from abnormalities of the brain that are due to genetically determined developmental malformations dating from early pregnancy or others caused by a variety of teratogenic insults. Cerebral palsy due to birth injury should also be distinguished from postnatal

Table 5–4. *Classification of Cerebral Palsy (Minear, 1956)**

Physiologic (motor)
- Spastic
- Athetotic
  - Tension
  - Nontension
  - Dystonic
  - Tremor
  - Rigidity
  - Ataxic
  - Tremor
  - Atonic (rare)
  - Mixed
  - Unclassified

Topographical
- Monoplegia
- Paraplegia
- Hemiplegia
- Triplegia
- Quadriplegia
- Diplegia
- Double hemiplegia

Etiologic
- Prenatal
  - Hereditary
  - Acquired in utero
- Natal
  - Anoxia
- Postnatal
  - Trauma (subdural hematoma, skull fractures, wounds, contusions of the brain)
  - Infections (meningitis, encephalitis, brain abscess)
  - Toxic causes (lead, arsenic, coal tar derivatives, streptomycin, etc.)
  - Vascular accidents
  - Anoxia (carbon monoxide poisoning, strangulation, high altitudes, deep pressure anoxia, hypoglycemia)
  - Neoplastic or late development defects (brain tumors, hydrocephalus, brain cysts, internal hydrocephalus)

Supplemental
- Psychological evaluation
  - Degree of mental deficiency, if any
- Physical status
  - Physical growth evaluation (Wetzel Grid or other)
  - Developmental level (Gesell)
  - Bone age
  - Contracture
- Convulsive seizures
- Posture and locomotive behavior patterns
- Eye-hand behavior patterns
- Visual status
  - Sensory
    - Amblyopia
    - Field defects
  - Motor
- Auditory status
  - Pitch range loss
  - Decibel loss
- Speech disturbances

Functional capacity (degree of severity)
- Class I. Patients with cerebral palsy with no practical limitation of activity
- Class II. Patients with cerebral palsy with slight to moderate limitation of activity
- Class III. Patients with cerebral palsy with moderate to great limitation of activity
- Class IV. Patients with cerebral palsy unable to carry on any useful physical activity

Therapeutic
- Class A. Patients with cerebral palsy not requiring treatment
- Class B. Patients with cerebral palsy who need minimal bracing and minimal therapy
- Class C. Patients with cerebral palsy who need bracing and apparatus, and the services of a cerebral palsy treatment team
- Class D. Patients with cerebral palsy limited to such a degree that they require long-term institutionalization and treatment

*Adapted from Minear, W. L.: A classification of cerebral palsy. Pediatrics, *18*:841, 1956.

Table 5–5. *Classification of Cerebral Palsy Patients by Types (Crothers and Paine, 1959)**

Spastic monoplegia		
Spastic hemiplegia		
Prenatal or natal	right	left
Postnatal	right	left
Spastic tetraplegia		
Symmetric		
Asymmetric		
Spastic triplegia		
Spastic paraplegia		
Extrapyramidal cerebral palsies, not mixed		
Kernicterus		
Mixed types		
Cerebral palsy plus cord injury		

*Adapted from Crothers, B., and Paine, R. S. The Natural History of Cerebral Palsy. Cambridge, Mass., Harvard University Press, 1959.

or acquired cerebral palsy, the consequence of insults sustained by the infant following birth.

Abnormalities of pregnancy, labor, and delivery may cause "hypoxic," "traumatic," or "toxic" damage to the brain.

Hypoxia. In the last trimester of pregnancy, probable causes of fetal hypoxia are: (1) antepartum hemorrhage due to placenta previa or other causes, with the attendant disturbance in placental nutrition; (2) pre-eclamptic toxemia—infarction of placentae of toxemic mothers tends to be more extensive than in mothers without toxemia; and there is greater decline of the oxygen saturation of the umbilical vein in pregnancy complicated by pre-eclampsia,

Table 5–6. *Classification of Cerebral Palsy in Childhood (Ingram, 1955; Balf and Ingram, 1956)**

Neurological Diagnosis	*Extent*	*Severity*
Hemiplegia	Right Left	Mild Moderately severe Severe
Bilateral hemiplegia		Mild Moderately severe Severe
Diplegia Hypotonic Dystonic Rigid or spastic	Paraplegic Triplegic Tetraplegic	Mild Moderately severe Severe
Ataxia	Unilateral Bilateral	Mild Moderately severe Severe
Dyskinesia Dystonic Choreoid Athetoid Tension Tremor	Monoplegic Hemiplegic Triplegic Tetraplegic	Mild Moderately severe Severe
Other		

*Adapted from Ingram, T. T. S.: A study of cerebral palsy in the childhood population of Edinburgh. Arch. Dis. Child., *30*:87, 1955, and Balf, C. L., and Ingram, T. T. S.: Problems in the classification of cerebral palsy in childhood. Brit. Med. J., *2*:163, 1955.

as compared with normal pregnancies;[182] (3) postmaturity; and (4) maternal causes of anoxemia such as cardiopulmonary disease.

Hypoxic damage to the fetus during labor and delivery may be caused by umbilical cord prolapse or torsion, or both, resulting in obstruction of circulation to the cord.

Neonatal apnea is not only the end result of many different forms of fetal injury, but is also a cause of further hypoxic damage in the newborn infant. The common causes of a failure to breathe after birth are prematurity and hypoxia during pregnancy or delivery. There may be poisoning or structural damage to the respiratory center, or the air passages may be obstructed as a result of the infant's premature efforts to breathe before delivery. Other neonatal complications that may cause hypoxia in the period immediately following birth are: persistent atelectasis due to immaturity, bronchial obstruction, or the baby's failure to expand the lungs; or hindrance of pulmonary respiratory exchange by hyaline membrane formation, pulmonary edema, intrauterine pneumonia, and aspiration of gastrointestinal contents.

Traumatic Birth. In recent years, the tendency has been to attribute less importance to trauma as an etiologic factor in birth injury and to emphasize the dangers of hypoxia. However, there is quite adequate evidence to suggest that subdural hemorrhage is predominantly the result of birth trauma. The forms of abnormal labor and delivery that are especially prone to cause subdural hemorrhage are: prolonged labor because of disproportion or malpresentation, precipitate delivery, forceps delivery, breech extraction, and version and extraction. Subdural hemorrhage most often results from tears of the dural ligaments that involve either the tributaries of the sagittal sinus or the great cerebral vein itself. Tears are especially liable to occur when undue or oblique stresses are placed on the tentorium cerebelli or the falx.[86]

Toxic Injury. Fetal damage may be caused by toxic agents that operate during late pregnancy, labor, and delivery. These include: (1) conditions that act by causing

toxic accumulations of naturally occurring substances (e.g., rhesus incompatibility producing an excess of bilirubin and ammonia in the fetus; maternal uremia, causing an excess of nitrogenous waste products to accumulate; or diabetes, in which an excess of a variety of hormonal substances that are damaging to the fetus are produced) or (2) the presence of abnormal toxins that cause fetal injury (as in syphilis and toxoplasmosis, in which secondary infection of the fetus is relatively common; or in some other maternal infection, e.g., pyelitis, diphtheria, or meningococcemia, in which there is no invasion of the fetus by microorganisms).

DEVELOPMENTAL MALFORMATIONS

Findings that suggest developmental malformations as a possible cause of cerebral palsy are: (1) known family history of cerebral palsy, congenital malformations, or neurologic disease (excluding mental retardation) in siblings, parents, uncles, aunts, or cousins; (2) births of patients after apparently uncomplicated pregnancy, labor, and delivery that were not thought to have been likely to cause gross hypoxia or trauma; (2) associated congenital malformations of patients, excluding those possibly secondary to cerebral palsy; (4) patients' having extremely small heads with an occipitofrontal circumference of less than one percentile for age.

The problem of etiologic diagnosis of developmental malformations as a cause of cerebral palsy is complex, and positive diagnostic criteria are difficult to establish. Clinically, developmental malformations might be responsible, in a significant proportion of patients, for bilateral hemiplegia and ataxic diplegia.

Experimental work and clinical observations have shown that a number of agents are likely to provoke developmental abnormalities in the unborn child, usually in the first three months of pregnancy. The connection between roentgen irradiation and congenital fetal defects is well established.[4,130,188] The most frequent neurologic manifestations are diplegia and ataxia, often complicated by epilepsy.[63] Rubella in the early months of pregnancy tends to produce offspring with congenital cataracts and abnormalities of other systems, with the brain being affected in a significantly high proportion of cases. Spastic paraplegia and athetosis may be the presenting clinical picture.

CAUSES OF ACQUIRED CEREBRAL PALSY

Well-recognized causes occurring in postnatal life include intracranial trauma, cerebral embolism, arterial thrombosis, intracranial abscess, venous thrombosis of the lateral sinus, meningitis, and viral encephalitides.

Perlstein and Barnett state that there is a statistically significant correlation between certain etiologic causes and specific clinical syndromes. In general, brain damage caused by anoxia is usually followed by extrapyramidal syndromes, whereas that caused by primary trauma and hemorrhage results in pyramidal affections. The following list shows some of the more common neurologic sequelae of various etiologic factors:[139]

Prematurity	spastic paraplegia
Breech delivery	athetoid or spastic paraplegia
Toxemia of pregnancy	spastic hemiplegia or quadriplegia
Birth trauma	spastic hemiplegia or quadriplegia
Anoxia	athetosis
Rh factor and kernicterus	athetosis with deafness and paralysis of supravergence
Maternal rubella	spasticity, with deafness or auditory aphasia, cataract, and congenital heart disease
Precipitate or cesarean delivery	spastic quadriplegia, ataxia, or rigidity
Placenta previa and abruptio	athetosis

Blumel, Eggers, and Evans, in a study of 100 cerebral palsy patients, reported the chief causes to be trauma at birth (13 per cent), anoxia (24 per cent), prematurity (32 per cent), congenital defects (11 per cent), and postnatal causes (7 per cent).[24] More than one agent may be operative in

producing cerebral palsy, and for the most part, the possible causes cannot be definitely identified.

In the past, the teaching has been dogmatic, it being said that spasticity is the result of damage to the motor cortex or pyramidal tracts, that athetosis is caused by lesions of the basal ganglia, that ataxia is due to damage or disease of the cerebellum or its connections, and that tremor and rigidity are the sequelae of more widespread lesions of the central nervous system. Recently, the gradual accumulation of more reliable and accurate information has failed to show a definite correlation between the pathologic findings and clinical features.[48] Generally, destructive, infectious, or vascular lesions will produce unilateral or asymmetrical paralysis, whereas developmental malformations will result in symmetrical involvement, although two or more destructive injuries could produce symmetrical paralysis.

Neurophysiologic Considerations

For a basic understanding of the musculoskeletal manifestations of brain damage in children, it is imperative to have a clear knowledge of the following fundamentals of the pathophysiology of the central nervous system.

SPASTICITY

Spasticity may be defined as a state of increase in tension of a muscle when it is passively lengthened, which is caused by an exaggeration of the muscle stretch reflex. It occurs in association with lesions of the cerebrum and descending pathways of the so-called pyramidal level of function. In the past, spasticity has been ascribed to the loss of, or release from the normal inhibiting action of, the pyramidal cortex on the anterior horn cells. Recent work indicates that spasticity results from an imbalance of the inhibitory and fasciculatory centers in the midbrain and brain stem reticular formations with consequent alteration of the alpha and gamma motor neuron balance.

Increased tension of a spastic muscle may be demonstrated on rapid or forceful passive movement; there is "blocking" and limitation of further movement. If passive movement of the limb is performed slowly, it is comparatively free. If the part is moved abruptly or suddenly, this "blocking" may be felt from the beginning of the passive movement, following which the muscle resists to a certain point and then relaxes. This "clasp knife" type of waxing and waning resistance of spasticity is distinguished from rigidity.

Objective means for accurate measurement of the degree of spasticity are not available. Thus far, clinical evaluation remains the most reliable method. The degree of hypertonicity and the range of motion of spastic limbs may vary between examinations and among different examiners. On palpation a spastic muscle may be hard, or it may be soft and flabby, depending not so much on the actual amount of spasticity as on the degree of contraction or relaxation at the time of palpation. The degree of electrical neuromuscular activity of a spastic muscle depends on whether the muscle is relaxed or stimulated.

In spasticity, the deep tendon reflexes are exaggerated and pathologic reflexes such as the Babinski and Hoffmann signs are present. On sudden dorsiflexion of the ankle or rapid distal movement of the patella, one may elicit *clonus*—alternate spasm and relaxation of the agonist and antagonist muscles.

Testing of motor power is difficult in spasticity, but it is important and an attempt should be made. In charting the muscle examination, both the motor power and the physiologic status should be designated. Motor power is graded by the standard accepted by the National Foundation for Infantile Paralysis, and the same abbreviations are used: zero, *0*; trace, *T*; poor, *P*; fair, *F*; good, *G*; and normal, *N*. For physiologic status, the author uses the following notations and abbreviations: *S* for spastic (stretch reflex); *H* for hypotonic; *0C* for cerebral zero (i.e., the patient has no voluntary control over the muscle); and *IN* for innervation normal. Table 5–7 shows various possible physiologic status and motor strength combinations of muscles in cerebral palsy patients.[174]

In the motor evaluation of a child with spasticity, the following pitfalls should be