The clinical picture for therapy and management

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Cerebral palsy is the commonly used name for a group of conditions characterised by motor dysfunction due to non-progressive brain damage early in life. There are usually associated disabilities as well as emotional, social and family difficulties. Cerebral palsies are the most common cause of childhood disability. The range of severity may be from total dependency and immobility to adequate abilities of talking, independent self-care and walking, running and other skills, although with some clumsy actions. A number of people with cerebral palsy are now able to benefit from mainstream education and further education. They participate more in various activities in society. These opportunities are assisted by legislation, advances in technology and changing attitudes in their society. Bax and Brown (2004) have given an overview of the cerebral palsies.

The motor dysfunction

The brain damage results in disorganised and delayed development of the neurological mechanisms of postural control, balance and movement. The muscles activated for these motor aspects are therefore inefficient and uncoordinated. Individuals have specific impairments such as hypertonicity or hypotonicity with weakness, abnormal patterns of muscle activation including excessive co-contractions. There are absent or poor isolated movements (poor selective motor control), abnormal postures and problems with manipulation. Besides neuromuscular impairments, the motor dysfunction has musculoskeletal problems. There are biomechanical difficulties resulting from both the neuromuscular dysfunction and musculoskeletal problems, which add to this complex picture.

The motor dysfunction changes with both growth and a child's development. Change also depends on how an individual uses his body. Physiotherapy positively contributes to body function. However, the brain damage is not progressive, though the motor behaviour changes. Musculoskeletal problems may increase in late childhood and adolescence needing physiotherapy input to minimise this.

What matters most to a child and his family is the overall functional delay and abnormal performance. Therapists need to address these daily functional difficulties together with a child and his parents or directly with an older person with cerebral palsy (see Chapters 2 and 7). Therapists will assess and assume which of the impairments and functional components are responsible for any functional disabilities. The associated impairments and disabilities below also influence the motor function. It is encouraging to know that functional limitations can be minimised even though basic impairments cannot strictly be cured.

There are different views as to which motor impairments are responsible for the total motor dysfunction and what correlation exists between them. Views also differ as to which impairments can be changed, and if not, when to make adaptations, including use of equipment, so that function can still take place. The underlying motor dyscontrol is controversial. This is not surprising, as not all the normal and abnormal neurological mechanisms are fully understood. There are also various ideas on biomechanics. Research continues on the basic dyscontrol and biomechanics.

The first edition of this book (Levitt 1977) presented a synthesis of valuable contributions from different therapy systems, some of which had been regarded as mutually exclusive. This synthesis or eclectic approach was further developed to include ideas from motor control and motor learning systems. The new edition of this book continues to synthesise current contributions from different approaches. As many colleagues are now not wedded to any one system of therapy, selections of their views are presented as well as those from each of the author's own studies and experience.

As a child does not 'move by neurophysiology alone', not only various ideas on learning motor control have been integrated into the general therapy framework, but the influence of the context of a child's function is given special consideration. This takes place in a child's home, school and community. A child learns best in a familiar environment and gains meaning for what is being achieved clinically. It is primarily the motivation of a child by people in these contexts and a child's own intrinsic motivation which have a profound impact on his or her achievement. In addition, consideration needs to be given to any environmental physical constraints and social attitudes which challenge a child and older person with cerebral palsy.

Associated impairments and disabilities

Brain damage in cerebral palsy may also be responsible for special sense defects of vision and hearing, abnormalities of speech and language, and aberrations of perception (Hall 1984; Neville 2000). Included in the perceptual defects are the *agnosias*. The agnosias are difficulties in recognising objects or symbols, even though sensation as such is not impaired, and the patient can prove by other means to know or have known what the object or symbol is. There may also be *dyspraxias*, some of which are also called visuomotor defects. This means that the child is unable to perform certain movements even though there is no paralysis, because the patterns or *engrams* have been lost or have not developed. Dyspraxia can involve movements of the limbs, face, eyes, tongue or be specifically restricted to such acts as writing, drawing and construction or even dressing. In other words, there seems to be a problem in 'motor planning' in those children who are dyspraxic. Some children may also have various behavioural problems such as distractibility and hyperkinesis, which are based on the brain damage. All these defects result in various learning problems and difficulties in communication. In addition, there may be intellectual impairment and various epilepsies (Himmelmann *et al.* 2006).

Not every child has some or all of these associated impairments. Even if the impairment were only motor, the resulting paucity of movement would prevent the child from fully exploring the environment. He is therefore limited in the acquisition of sensations and perceptions of everyday things. A child may then *appear* to have defects of perception, but these may not be due to the brain damage but caused by lack of experience. The same lack of everyday experiences retards the development of language and affects the child's speech. His general understanding may suffer, so he appears to be intellectually retarded. This can go so far that normal intelligence has been camouflaged by severe physical disability. Furthermore, the lack of movement can affect the general behaviour of the child. Thus, some abnormal behaviour may be due to the lack of satisfying emotional and social experiences for which movement is necessary. Motor dysfunction may therefore interact with emotional and social development of a child. However, positive attitudes in a family and child can encourage optimum development.

Teamwork. It is therefore important for any therapist to recognise that motor function cannot be isolated from other functions and that she is treating a child who is not solely physically but multiply disabled. Therapists will also need to consider when the associated physical and behavioural problems constrain motor function (Thylefors *et al.* 2000).

In order to manage the multiple disabilities and lack of related learning experiences which interfere

with a child's development, a physiotherapist or occupational therapist needs to be part of a team. The teamwork varies in different places such as community centres, child development centres, units in hospitals or within educational settings. Teamwork is discussed in Chapters 2, 8, 10 and 12.

Aetiology

Premature infants are at greater risk of brain dysfunction. There are many causes of the brain damage, including abnormal development of the brain, anoxia, intracranial bleeding, excessive neonatal asphyxia (hypoxic ischaemic neonatal encephalopathy), trauma, hypoglycaemia, anoxia as in near-drowning, choking, neurotrophic virus and from various infections. These have been extensively discussed in the medical literature (Rosenbloom 1995; Hagberg *et al*. 1996; Stanley *et al*. 2000; Himmelmann *et al*. 2005). The therapist is, however, rarely guided by the aetiology in her treatment planning. In some cases the cause is not certain, and in many cases knowing the cause does not necessarily indicate a specific diagnosis or specific treatment. Nevertheless, the therapist should acquaint herself with the history of the case. Many of these children have been affected from infancy and have been difficult to feed and handle. Many hospitalisations and separations of babies from parents may happen in the early period. This may easily have influenced the parent–child relationships so essential for child development. Furthermore, the history may sometimes give an indication of the prognosis; for example, with marked microcephaly with severe multiple impairments the prognosis would be poor.

Clinical picture and development

It is important to recognise that the causes of cerebral palsy take place in the prenatal, perinatal and postnatal periods. In all cases, it is an immature nervous system which suffers the insult and the nervous system afterwards continues to develop in the presence of the damage. The therapist must therefore not think of herself as treating an upper motor neurone lesion in a 'little adult' nor can she regard the problem solely as one of retardation in development. What the therapist faces is a complex situation of pathological symptoms within the context of a developing child (Sheridan 1975, 1977; Drillien & Drummond 1977, 1983; Illingworth 1983; McGraw 1989; Sheridan *et al*. 2008). There are six main aspects to the clinical picture:

(1) Retardation in the development of new skills expected at the child's chronological age.
(2) Persistence of infantile behaviour in all functions, including infantile reflex reactions.
(3) Slow rate of progress from one developmental stage to the next.
(4) A smaller variety of skills than in the able-bodied child.
(5) Variations in the normal sequence of skills.
(6) Abnormal and unusual performance of skills.

In order to recognise abnormal motor and general behaviour, the therapist should know what a normal child does and how he does it at the various stages of his development. Information on each individual child's developmental levels should be sought from the consultants and other members of the cerebral palsy team. Reference will have to be made to the extensive literature on the field of child development.

Although normal child development is the basis on which the abnormal development is appreciated, it does not follow that assessment and treatment should rely upon a strict adherence to normal developmental schedules. Even 'normal' children show many variations from the 'normal' developmental sequences and patterns of development which have been derived from the *average* child. Cultural differences exist for normal motor development (Solomons & Solomons 1975; Hopkins & Westra 1989). However, in any culture, the child with cerebral palsy will show additional variations due to neurological and mechanical difficulties. If one considers, say, the normal developmental scales of gross motor development, in cerebral palsy a child has frequently achieved abilities (components) and motor functions at one level of development, omitted abilities at another level and only partially achieved motor abilities and functions at still other levels. There is thus more of a scatter of abilities and whole motor functions than in able-bodied children. The analysis of motor function into components is discussed in Chapters 5, 6, 8 and 9.

If the gross motor development is generally considered to be around a given age, the development of hand function, speech and language, social and emotional and intellectual levels may all be at different ages. None of these ages may necessarily coincide with the child's chronological age.

Therefore, the developmental schedules in normal child development should only be used as *guidelines* in treatment, and adaptation should be made for each child's disabilities and individuality (see Chapter 9).

More attention is usually given to motor development rather than other avenues of development, as it is the motor dysfunction which characterises cerebral palsy. Here again, the therapist should remember that abnormal motor behaviour interacts with other functions. Each area of development – such as gross motor, manipulation, speech and language, perception, social and emotional adjustments, and cognition – interacts as well as has its own pattern or avenue of development. Furthermore, the potential for function is dependent not only on the disabilities present but also on a child, his personality and 'drive' as well as his capacity to learn. Therefore, a total habilitation programme is necessary and should be planned to deal with the whole development of each child.

Whilst aiming at the maximum function possible, the therapists concerned must take account of the *damaged* nervous system and adjust their expectations of achievements by the child. This depends on a therapist's clinical experience as prognosis is difficult in view of the multiple factors involved. There are measures of the severity of a child's disability in Chapter 8, which guide the expectations of a therapist, but overdependence on levels of severity may not always be reliable in individual children.

Change in clinical picture

As the lesion is in a developing nervous system, the clinical picture is clearly not a static set of signs and symptoms for treatment. But whilst the lesion itself is non-progressive, its manifestations change as the nervous and musculoskeletal systems mature. As more is demanded of the child, the degree of the motor disability appears to be greater. For example, a 3-year-old is expected to do more than a baby, and therefore his difficulties are greater for the same pathology.

In addition, the pathological symptoms may develop with the years. Spasticity may increase, involuntary movements may only appear at the age of 2 or 3 years and ataxia may only be diagnosed when the child walks or when grasp is expected to become more accurate. Diagnoses may change as the baby develops to childhood, and especially as the child becomes more active. For example, a monoplegia reveals itself as a hemiplegia. Later a triplegia reveals itself as a tetraplegia. Cerebral palsies have an emerging diagnosis. Later, especially in adolescence, growth and increase in weight contribute to apparent deterioration. Recent research identifies that deterioration is not inevitable in all cases (see Chapter 7).

Treatment and management in infancy. The earlier the treatment is started, the more opportunity is given for whatever potential there may be for developing any normal abilities and for decreasing the abnormal movement patterns and postural difficulties (Kong 1987; de Groot 1993). However, abnormalities detected in infants may be transient as some infants overcome them without intervention. Therapists offer pleasurable and a variety of developmentally appropriate and active motor activities enjoyed by both parent and baby. During intervention, therapists observe if a baby or young child makes his own efforts to move using compensatory or adaptive patterns which can be 'good enough' but block the development of more efficient patterns or result in 'learned disuse' of a body part. Any immobility threatens musculoskeletal growth and development which can lead to deformities. Early physiotherapy minimises such problems.

The value of early developmental intervention is to provide an increase in a baby's sensory-motor and everyday experiences and interaction with his mother and father. The sooner a baby can be helped to move, the sooner he can explore and the sooner he can communicate the information he gains through such exploration. The therapist is in fact contributing to his learning and understanding as well as enabling him to bond with his mother and father.

Although the clinical picture is known to change with the years, it is not yet possible to predict the natural history of the condition in each particular child. Infants and babies with marked early neurological signs may later prove to be only mildly affected, or even normal (Ellenberg & Nelson 1981; Nelson & Ellenberg 1982). On the other hand, apparently mildly affected ones may become

progressively worse with the years. It is therefore difficult to prove the value of a number of different early treatment approaches (Vojta 1984; Kong 1987; Katona 1989; Morris 1996). However, research in neonatal physiotherapy continues. Blauw-Hospers and Hadders-Algra (2005) have found positive effects on babies at term, rather than preterm, with specific and general developmental early treatments in their systematic review of 12 studies. The review by Spittle *et al.* (2007) found little evidence of early intervention on motor development. Reviews point out that the studies involve heterogeneous samples.

Nevertheless, until we know more definitely which babies are going to 'come right' on their own, it is better to let them have the benefit of treatment so that any potentials for improvement are not lost. Despite the controversy as to the value of early treatment, there is clearly no doubt about its importance to the parents, who receive a great deal of practical advice and support from the therapists. Among others, Goodman *et al.* (1991) found that if their research could not firmly state that neonatal physiotherapy was responsible for babies' motor developmental progress, all mothers confirmed their great appreciation for the support and practical ideas from their physiotherapists. Olow (1986) emphasises that early intervention reduces the frustration of early rearing of children with disabilities. Whilst medical practitioners are watching the development of the child in order to make a reliable diagnosis, the parents have to live with that child throughout each day of those months and years. Parents need support and practical ideas for feeding, childcare and motor activities for their child throughout the emerging diagnoses. This is an essential part of the therapist's management programme with them. Well-supported parents are most likely to benefit their young children's development (see Chapter 2).

Treatment and management in childhood, adolescence and adulthood. During these changes in the clinical picture, treatment and management programmes need to relate to an individual's wider environments of the playgroup, nursery, preschool, schools, adult day care centres and work places. The persons with cerebral palsy at different ages also change through interaction with the variety of personnel in environments in which they find themselves. Physiotherapy and occupational therapy as well as other therapies are therefore being planned across a lifespan of each person with cerebral palsy.

Management will include working with orthotists, orthopaedic surgeons and other consultants. The therapist will share selected skills and advice on equipment with anybody closely involved with each person having a disability.

Classification

Numerous classifications and subclassifications have been proposed by different authorities, and though clinically helpful, none of these diagnostic labels suffice to formulate adequate treatment plans. The therapist must also have a detailed assessment based primarily on motor functions in order to work out a treatment programme.

Classifications of topography of cerebral palsy

The topographical classifications frequently used are as follows:

Tetraplegia (quadriplegia). Involvement of all limbs and body. Arms are equally or more affected than the legs. Many are asymmetrical (one side more affected).

Diplegia. Involvement of limbs, with arms much less affected than legs. Asymmetry may be present.

Hemiplegia. Limbs and body on one side are affected.

Neville and Goodman (2001) present different authors in a book on congenital hemiplegia. These topographical classifications can be imprecise, as they may change with a child's development. One useful upper limb may convey a triplegia which could become a tetraplegia. Upper limbs may appear unaffected, suggesting a paraplegia but being really a diplegia with only fine-hand use being affected when this is later expected. Hemiplegia may have minor involvement on the unaffected side. A monoplegia is rare, usually becoming a hemiplegia with increased activity.

Classification of types of cerebral palsies

There are spastic types, athetoid (dyskinetic) types and a rare ataxic type. There is a hypotonic type

which either becomes a spastic, athetoid or ataxic type. There is a transient dystonic stage in babies before they are diagnosed as a spastic or dyskinetic type of cerebral palsy (Bax & Brown 2004). Tetraplegias usually have either spasticity, dystonia, dyskinesia (athetosis), hypotonia or ataxia. Hemiplegia is usually a spastic type often starting out hypotonic. Hemi-athetoids with or without dystonia are occasionally seen. Once again, classifications are not always clear-cut and the therapist may have to treat impairments of one type in another type. The predominant impairments will contribute to the diagnostic type referred for therapy. Developmental functional training is nevertheless indicated for all types of cerebral palsies.

Spastic cerebral palsy

Main motor characteristics are as follows:

Hypertonus. If spastic muscles are stretched at a particular speed, they respond in an exaggerated fashion. They contract, blocking the movement. If this sudden passive stretch is continued, the spasticity may melt away in some cases. The movement block is the 'catch' and with the subsequent movement this is called a 'clasp-knife' variety of spastic hypertonus. This hyperactive stretch reflex may occur at the beginning, middle or near the end of the range of movement. There are increased tendon jerks, occasional clonus and other signs of upper motor neurone lesion. The velocity-dependent hyperactive stretch reflex is the physiological definition of spasticity. Stiffness is not true spasticity and may or may not accompany the reflex reaction to brisk passive stretch. Viscoelastic muscle and soft tissue changes are also causes of stiffness (Katz & Rymer 1989; Dietz & Berger 1995). However, clinicians usually use 'spasticity' and 'spastic muscles' as an umbrella term for stiffness of limbs and recognise that other motor symptoms are also included under this umbrella. These are discussed below. Current views are that the hyperactive stretch reflex is not as much the cause of abnormal function as weakness (Lin 2004; Ross & Engsberg 2007). Movements are usually slower than the velocity needed to obtain the hyperactive stretch reflex.

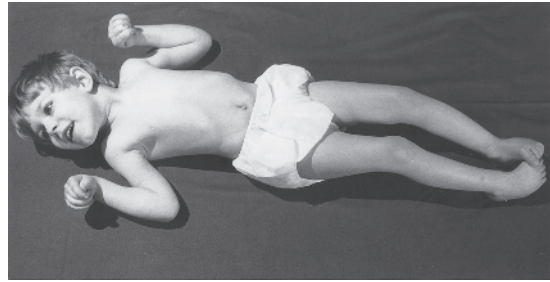Hypertonus may be either spasticity or rigidity (dystonia). The overlap between the two is almost

Figure 1.1   Child with spastic quadriplegia. Head preference to right, shoulders protracted, semi-abduction, elbows flexed-pronated, wrists and fingers flexed, thumb adducted. Hips and knees flexed, tendency to internal rotation–adduction with feet in equinovarus, toes flexed.

impossible to differentiate when severe. A mixture of spasticity and rigidity may be diagnosed (Lin 2004). Rigidity is recognised by a *plastic* or continuous resistance to passive stretch throughout the full range of motion. This *lead-pipe* rigidity differs from spasticity as spasticity offers resistance at a point or small part of the passive range of motion. Spasticity is selective affecting specific muscles, for example giving a predominantly flexor pattern in the arm and extensor pattern in the leg. Rigidity (dystonia) affects all muscle groups equally. Drugs such as botulinum toxin A, oral and intrathecal baclofen are used to control spasticity and dystonia (Lin 2004), together with a physiotherapy programme.

Abnormal postures (see Figs 1.1–1.3). These are usually associated with the antigravity muscles which are extensors in the leg and the flexors in the arm. However, the therapist will find *many* variations on this, especially when the child reaches different levels of development (Bobath & Bobath
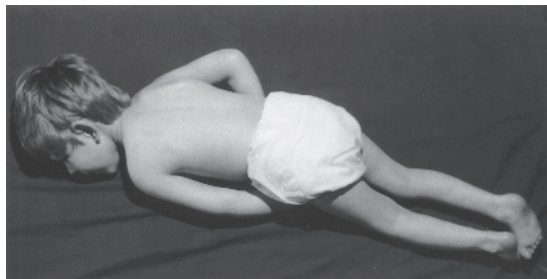

Figure 1.2   Same child with quadriplegia with postural changes in prone. Asymmetry of arms caught under body. Hips and knees flexed, feet in equinovarus. Head preference is now to left.

Figure 1.3 Same child being taught to sit by his father. Head preference to right, shoulders protracted, elbows flexed-pronated, hands flexed, knees and feet held symmetrical with hips. Symmetrical trunk.

1972). Children have floppiness of the head and trunk together with stiff spastic limbs. This is associated with delayed development of the mechanisms of postural stabilisation and postural adjustment of the head and trunk as well as delay in stabilisation of pelvic and shoulder girdles (Foley 1977, 1998). The postural mechanisms and the common abnormal postures in supine, prone, sitting, standing and hand positions are described in Chapters 5, 9 and 11.

The abnormal limb postures become held by stiff shorter 'spastic' muscle groups whose lengthened antagonists are weak, or apparently weak, in that they cannot overcome the tight pull of the shortened muscles and so correct the abnormal postures. The short muscle groups are also weak and cannot easily assume a normal alignment of posture. In time, there is transformation of structure of the stiff spastic muscle (see below). Besides spasticity, there are therefore various other causes of abnormal postures of limbs and body including the weakness and especially compensation for absent or poor postural mechanisms of balance control. Spasticity is therefore not overemphasised as a cause in this and previous editions of this book.

Abnormal postures appear as unfixed deformities, which in time may become fixed deformities or contractures with subsequent bony deformities.

Abnormal postures and deformities, particularly in the upright positions, contribute to abnormal gaits.

Changes in spasticity and postures. These changes may occur with excitement, fear or anxiety, and pain, which increases muscle tension. Shifts in spasticity may occur in the same affected parts of the body or from one part of the body to another in, say, stimulation of abnormal reactions such as occasional remnants of tonic reflex activity. Changes in spasticity are seen with changes of position in some children. Position of the head and neck may affect the distribution of spasticity. Sudden or fast movements, rather than slow movements, increase spasticity.

Voluntary movement. Spasticity does not necessarily mean paralysis. Voluntary motion is present and may be laboured. There may be weakness in the initiation of motion or during movement at different parts of its range. If spasticity is decreased or removed by treatment or drugs, the spastic muscles may be found to be weak. For example, the removal of spasticity of the gastrocnemius with botulinum toxin A injection reveals weak plantarflexion. Spastic muscles may have specific structural changes due to adaptability to abnormal use or disuse (Tabary *et al*. 1981). Initially, spastic muscles are, however, *structurally* normal though not normally extensible (Tardieu *et al*. 1982). Therefore, spastic muscles tend to shorten in dynamic deformity and later may become fixed contractures. Once spasticity is decreased the antagonists may also be stronger once they no longer have to overcome the resistance of tight spastic muscles and can work in mid-range or full range. However, in time these antagonists may have become weak with disuse within the muscle imbalance between agonists and antagonists.

The groups of muscles or *chains* of muscles used in the movement patterns (muscle activation patterns) are different from those used in normal children of the same age. Either the muscles which work in association with each other are stereotyped and are occasionally seen in the normal child, usually at an infantile level of movement, or the association of muscles is abnormal. For example, hip extension–adduction–internal rotation is normally used in creeping movements or within the push-off in walking but many other combinations must be used during the full execution of creeping and walking. This may be impossible and a child only uses the same pattern at all times in the motor skill. One example of a normal arm pattern is

shoulder flexion–adduction with some external rotation for feeding or combing one's hair. In the case of the child with spasticity, the arm pattern is usually flexion–adduction with *internal* rotation and *pronation* of the elbow. The ability to fractionate movement is very difficult for the child, for example to maintain flexion at the shoulder and extension of the elbow and wrist when reaching for an object. The arm pattern usually tends to persist in flexion at all joints.

Co-contraction of the agonist with the antagonist instead of the normal reciprocal relaxation persists in the spastic type of cerebral palsy. Normal co-contraction is also evident in any person attempting a new and difficult skill in hand function or in the legs. Before the postural control develops in normal infants there is a co-contraction response in weight-bearing and co-contraction features in early stages of walking in children without cerebral palsy. These patterns persist in cerebral palsy (Leonard *et al.* 1991; Foley 1998; Lin 2000). The co-contraction provides some stability but for a more flexible mature gait, postural control training is essential. Voluntary arm and leg movements are also directly affected by poor postural control, as this interferes with their efficiency creating weakness of both postural muscles and voluntary synergies (movement patterns).

Lack of isolated or discrete movements (selective motor control) and fine motor coordination are also delayed in younger able-bodied children as well as in the spastic type, particularly if severe.

Associated impairments

(1) Intelligence varies and is usually more impaired in tetraplegia.
(2) Sensory loss occasionally occurs in hemiplegia with visual field loss and lack of sensation in the hand (Tizard *et al.* 1954). Sensory dysfunction such as sensory discrimination and sensory integration rather more than sensory loss is present in individuals (Lesny *et al.* 1993; Yekutiel *et al.* 1994). Lack of sensory awareness and sensory information for motor actions often relates to poor motor experience rather than loss of sensation. A child may be hyposensitive or hypersensitive to sensory input, so sensory-motor therapy needs to be carefully assessed.
(3) Perceptual problems especially of body and spatial relationships are more common in the spastic type of cerebral palsy. They relate to sensory dysfunction and cognitive problems as well as to poor sensory-motor experiences.
(4) Poor respiration with later rib cage abnormalities may exist.
(5) Feeding problems exist, particularly in tetraplegia.
(6) Growth of hemiplegic limbs or severely affected lower limbs in bilateral cases can be less than the other limbs.
(7) Epilepsies are more common in tetraplegia and hemiplegia but minimal in diplegia (Neville 2000).
(8) A congenital suprabulbar palsy is found in some tetraplegias with mild spasticity (Neville 2000) or severe involvement.

Athetoid (dyskinetic, dystonic) cerebral palsy

Main motor characteristics are as follows:

Involuntary movements – athetosis. These are bizarre, purposeless movements which may be uncontrollable. The involuntary movements may be slow or fast; they may be writhing, jerky, tremor, swiping or rotary patterns or they may be unpatterned. They are present at rest in some children. The involuntary motion is increased by excitement, any form of insecurity and the effort to make a voluntary movement or even to tackle a mental problem. Factors which decrease dyskinesia (athetosis) are fatigue, drowsiness, sleep, fever, prone lying or the child's attention being deeply held. Involuntary motion may be present in all parts of the body including the face and tongue. Dyskinesia may only appear in hands or feet or in proximal joints or in both distal and proximal joints. Generally the child finds great difficulty in being still.

Postural control. The involuntary movements or dystonic spasms may throw a child off balance. However, the well-known instability in children with dyskinesia is often directly connected with the postural mechanisms discussed in Chapter 5 (Foley 1983). Foley (1998) relates involuntary motion with abnormal tilt reactions. Abnormal standing postures usually involve backward lean with hip extension, lordosis and kyphosis with chin jutting well forward. This is a compensation for instability.

Voluntary movements. These are possible but there may be an initial delay before the movement is begun. The involuntary movement may partially or totally disrupt the willed movement, making it uncoordinated. There is a lack of finer movements and weakness. Grasp and release have extreme flexion and extension movements which some older children learn to control for finer grasp or use of large keys on a computer.

Hypertonia or hypotonia. Either they may exist or there may be fluctuations of tone. The hypertonus or dystonia is a 'lead-pipe' or 'cog-wheel' rigidity. There is a continuous resistance to passive stretch throughout full range of motion. Dystonia can be particularly disabling, especially if combined with spasticity. Arousal of emotions increases tone. Sudden flexion or extensor spasms could occur. Sudden wide opening of the mouth with spasm can take place. Sleep decreases spasms or dystonic postures. Deformities are less likely due to the fluctuations in muscle spasms and stiffness.

The athetoid dance. Some athetoids are unable to maintain weight on their feet and continually withdraw their feet either upwards, or upwards and outwards, in an 'athetoid dance'. They may take weight on one foot whilst pawing or scraping the ground in a withdrawal motion with the other leg. This has been attributed to a conflict between grasp and withdrawal reflexes. This conflict of reflexes may also be seen in the hands (Twitchell 1961). A common pattern is a 'run headlong' using momentum as they cannot stand still nor adjust their posture for slower walking. They run before they can walk.

Paralysis of gaze movements may occur, so athetoids may find it difficult to look upwards and sometimes also to close their eyes voluntarily. Poor head control also disrupts use of the eyes.

The dyskinetic types change with time. They may be floppy in babyhood and only exhibit the involuntary movements when they reach 2 or 3 years of age. Adult athetoids do not appear hypotonic but have muscle tension. Muscle tension also seems to be increased in an effort to control involuntary movements. The standing posture of late childhood, adolescence and adulthood is usually with extended hips, bent knees and pronated feet and rounded back with arms and chin held forward to counter the extension backwards (Fig. 1.4).
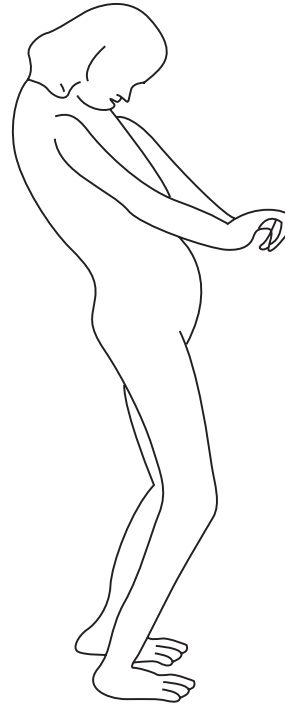

Figure 1.4 Person with dystonia/dyskinesia in standing or walking positions.

Associated impairments

(1) Intelligence is frequently good and may be very high. Intellectual impairment is occasionally present.
(2) Hearing loss of a specific high frequency type is associated with athetoids caused by kernicterus, though it is now a rare cause.
(3) 'Drive' and outgoing personalities are often observed. Emotional lability is more frequent than in other types of cerebral palsies.
(4) Articulatory speech difficulties and breathing problems may be present, and the child's oromotor problems create feeding difficulties. Poor arm function may adversely affect the development of self-feeding.

Ataxic cerebral palsy

Main motor characteristics are as follows:

Disturbances of balance. There is poor stabilisation of the head, trunk, shoulder and pelvic girdles.

Some ataxics overcompensate for this instability by having excessive balance-saving reactions in the arms. Instability is also found in children with any type of cerebral palsy and may be called ataxia in the dyskinetic or spastic type as pure ataxia is fairly rare. Unsteady gaits arise from the brain lesion affecting motor control (Foley 1998; Neville 2000).

Voluntary movements. They are present but clumsy or uncoordinated. The child overreaches or underreaches for an object and is said to have 'dysmetria'. This inaccurate limb movement in relation to its objective may also be accompanied by intention tremor. Poor fine hand movements occur.

Hypotonia. It is usual. There is excessive flexibility of joints and poor muscle power.

Nystagmus. It may exist.

Associated impairments

(1) Intellectual impairment may exist, especially in the presence of visual and perceptual problems.
(2) 'Clumsy' intelligent children are sometimes diagnosed as having ataxic cerebral palsy.
(3) A 'pure' ataxic is rarely diagnosed except for a group of genetic origin called 'dysequilibrium syndrome' (Neville 2000).

Common features in all types of cerebral palsies

Postural mechanisms

The classification into types of cerebral palsies has tended to obscure the fact that there are important motor features which are common to all types. For instance, all cerebral palsied children are delayed in motor development. However, the symptoms of the different types of cerebral palsies, such as spasticity, sudden spasms and the various involuntary movements, only play a part in this disturbance of development. Delayed or abnormal development of the postural balance mechanisms significantly disturbs the motor development. Postural mechanisms are an intrinsic part of motor skills. When they are absent or abnormal, this leads to absent or abnormal motor skills.

Chapters 5 and 9 discuss these aspects in detail, as they are fundamental to the framework for therapy.

A common feature is also associated weakness of neck, trunk, shoulder and pelvic muscles, which are not activated by undeveloped postural mechanisms.

Classification based on motor function

A classification based on motor function incorporates postural control, which is intrinsic to motor developmental functions. This is not directly based on any diagnostic type of cerebral palsy.

The Gross Motor Function Classification System (GMFCS) for children with cerebral palsy (Palisano *et al*. 1997, updated 2008) classifies children according to what they can do at different ages. There are five levels of classification, giving distinctions in self-initiated motor functions. Level I children function without restriction, only having limitations in advanced motor skills. The motor functions decrease from I to V, with level V representing children with severe motor restrictions. This is a clinical and research classification which is detailed. This classification provides good communication between colleagues internationally rather than 'mild, moderate or severe' classifications.

Abnormal reflexes

Besides the desirable postural mechanisms, there are abnormal reflexes which have no predilection for any specific type of cerebral palsy. These are infantile (primitive) reflexes which are present in the normal newborn and which become integrated or disappear as the baby matures. In cerebral palsied children, infantile reflexes are still present long after the ages when they should have become integrated within the nervous system. As children with cerebral palsy have not been able to develop more mature neurological postural mechanisms, the infantile reflexes can be their only way to function. Whilst there are many infantile reflexes, those of most interest to the therapist are the Moro reflex, the palmar and plantar grasp reflexes, automatic stepping, excessive neck righting reflex, positive supporting, extensor thrust and feeding reflexes (Capute *et al*. 1984; see Table 8.3). These reactions may be stimulated by either peripheral or cortical activations. Some children with severe multiple disabilities activate some of these reflex responses in their efforts

to balance, move or communicate non-verbally. A therapist needs to include knowledge of how her peripheral stimulation and handling might cause undesirable reflex responses instead of developing more advanced motor control. Examples of the use of reflexes are as follows: a child may use grasp reflexes to hold a small object, a plantar grasp to grip the floor for stability, automatic stepping when the body is fully supported in a walker and positive supporting reaction for standing in a standing frame. Children use extensor thrusts or Moro reactions to communicate non-verbally.

There are also the tonic reflexes, which are the tonic labyrinthine reflexes, the asymmetrical tonic neck reflex and the symmetrical tonic neck reflexes. Some neurologists group these tonic reflexes amongst the infantile reflexes, whereas others argue that they are not present in the normal infant and are always pathological. Tonic reflexes are only seen in the most severely impaired children (Foley 1977), especially if obligatory. These tonic reflexes are sometimes called *postural reflexes* but they are *abnormal* postural reflexes and should not be confused with the normal postural mechanisms as described by Rushworth (1961), Martin (1967), Foley (1977, 1998), Shumway-Cook and Woollacott (2001) and others.

The principle of treatment which therapists should follow in relation to the complicated collection of reflexes is *not* to go 'reflex hunting'. In the past, some therapists observed that reflexes interfere with motor function and speech. This does *not* always occur. The approach is to examine the function of the child first and, only when abnormality has been detected, to then consider whether *one* of the reasons for this abnormality seems to be a pathological or primitive reflex. However, it is the therapist's work in building a child's function that simultaneously can modify or overcome any reflex reactions in a child. Table 8.3 of primitive and tonic reflex reactions is given so that a therapist recognises any total or remnants of these reflex reactions or infantile (primitive) responses in individual children so that she can assess if children are using these infantile patterns as compensation for lack of motor control.

Recent research calls into question the importance of primitive (infantile) reflexes. They are no longer considered a substrate for motor control and are not reliable predictors of future motor development. New ideas on theoretical bases of motor training

disagree with therapy using the 'hierarchical lists' of primitive and tonic reflexes followed by more mature reactions (Cioni *et al.* 1989, 1992; Horak 1992; Prechtl 2001; Einspieler *et al.* 2005). These studies on reflexes lend support to 'avoid reflex hunting' expressed in this book since the first edition in 1977.

Additional impairments

Individual children, particularly with severe cerebral palsy, may have sleep problems, fatigue, feeding problems and poor nutrition, decreased bone mineral density, musculoskeletal pain or pain from severe gastroesophageal reflux, and are less fit than able-bodied children. Most of these problems develop in later childhood and are managed by medical consultants. Nevertheless, a therapist needs to be aware of them as they may impinge on the amount of energy a child has available for therapy programmes. Parents are often short of sleep as they need to comfort, feed or give medicines to their child at night. This impacts on their capacities to carry out a child's home therapy. Fitness is naturally in the realm of therapy and management. Pain and decreased bone mineral density are prevented to some degree by therapists using activities and weight-bearing postures.

Motor delay

Cerebral palsy consists of both motor delay and motor disorder. There are many other conditions which present similar problems of motor delay or of delay and disorder. All these conditions are also called the *developmental disabilities* (Pearson & Williams 1972; Levitt 1984).

They may be due to the following:

Intellectual impairment, which is caused by various metabolic disorders, chromosome anomalies, leucodystrophies, microcephaly and other abnormalities of the skull and brain, endocrine disorders and the causes of brain damage given for the cerebral palsies. Down's syndrome also creates motor delay.

Deprivation of normal stimulation associated with social, economic and emotional problems, including maternal depression.

Malnutrition alone, but usually together with deprived environments. Once malnutrition is treated, lack of normal stimulation may still retard the child's development.

The presence of non-motor impairments, which may lead to motor delay, for example severe visual impairments, severe perceptual defects, apraxias as well as intellectual disabilities mentioned above. Children with delay in any developmental area may have an associated delay in motor development (see the section on motor development and the visually impaired child in Chapter 9).

Presence of motor impairments other than the cerebral palsies. For example, spina bifida, the myopathies, myelopathies and various progressive neurological diseases and congenital deformities may obviously delay development of fine and gross motor function (Holt 1975).

Principles of treatment and organisation of treatment will be similar to those discussed in Chapters 2, 5, 6 and 9. *Specific problems* in the conditions above are considered in other publications (Levitt 1984; Eckersley 1993; Shepherd 1995; Burns & MacDonald 1996; Campbell *et al*. 2006; Tecklin 2008).

Principles of learning and therapy

Broad framework for therapy and management

A broad framework for therapy and management assists interpreting assessments and comprehensive programme planning for intervention. Details of how each aspect is implemented are not given. The World Health Organization's current International Classification of Functioning, Disability and Health (ICF) (WHO 2001) describes a person's functioning in terms of body structures, body functions, activity and participation.

Definitions of components in the ICF

Body functions are physiological functions of body systems (including psychological functions).

Body structures are anatomical parts of the body and limbs.

Impairments are problems in body function or structure such as abnormal balance, and deformity.

Activity is the execution of a task or action by an individual such as standing, walking, grasping. This can include daily living tasks such as eating, dressing, toileting and washing. However, these tasks can also be participation in life situations.

Participation is involvement in a life situation such as participation in an individual's community, as in some school activities, shopping, caring for children, social and sporting activities, and use of playgrounds.

Personal factors influence how disability is experienced by an individual. These include age, coping styles, character and overall behaviour.

Environmental factors affect the individual's function and participation. These include family and social attitudes, architectural barriers, climate and terrain.

Practical application

The components of the ICF model are not sequential. Participation in society may not depend on improving impairments when a person's own functional strategies are used and when special equipment, electric wheelchairs, computers and other technology are chosen. Depending on the severity of cerebral palsy, innovative functional strategies or motor compensations may allow independent function without focusing on impairments, for example MOVE (Bidabe & Lollar 1990) and Conductive Education (Hari & Akos 1988). Research by Charles *et al*. (2006) did not change impairment but improved function in the arm and hand. Quality of life is particularly dependent on participation. Bjornson *et al*. (2008), in their research with people aged 10 and over, found that 'functional level and performance did not influence quality of life'.

There are three points supporting this view confirmed by clinical experience:

- Owing to the brain damage, not all impairments can be minimised.
- When selected impairments have been minimised, this did not always carry over into daily function.
- Participation may not be dependent on impairments nor functional performance.

However, secondary impairments such as contractures and musculoskeletal pain may result from particular motor compensations, and impairments such as specific weakness, poor balance, abnormal

coordination and hypertonus may increase with time. These secondary impairments have limited the reliability of daily functions and limited the range of participation in an individual's home and community. Current and future research will clarify these different views for individuals with cerebral palsy. The relationships of impairment, activity and participation are complex. We cannot firmly maintain that treatment of impairment leads to function and that improvement of function leads to participation in the individual's different environments.

Therefore, therapy principles include:

- Assessment and management based on the perspectives of an individual, the family, teachers and others involved with that individual.
- Assessment and management of impairments which constrain functions and daily tasks needed.
- Assessment and prevention of secondary and increasing impairments.
- Focus on functional therapy and correction of impairments within function.
- Assess and manage function in the context of a person's home, school and community.
- Consideration of attitudes in family and society that disable a person.
- Encouragement of the personal attributes of an individual with disability and of his or her family.

There is increasing research on the relationships between the items in the ICF model, which will be discussed in this book.

Aims of physiotherapy, occupational therapy and speech therapy are:

(1) To develop forms of communication (gesture, speech, typing and alternative forms of communication with signs or electronic aids).
(2) To develop independence in the daily activities of eating, drinking, dressing, washing, toileting and general self-care with and without aids, such as special utensils, toys and special furniture.
(3) To develop abilities to play and achieve hobbies and recreational activities with or without adapted equipment.
(4) To develop some form of locomotion and independent mobility, which may include wheelchairs, playthings, tricycles or driving adapted motor vehicles.

All these aims need to be considered in terms of learning processes interacting with neurological and orthopaedic aspects and environmental constraints. Therefore all therapists draw on the fields of education and psychology and gain much from close teamwork with teachers, psychologists, social workers and psychotherapists. The psychotherapists and social workers are important as learning is intimately involved with emotions. Some learning models do not give adequate attention to this fact. The role of cultural factors in planning the programme needs to be considered by everyone. A collaborative learning approach initially developed by the author in consultation with a psychiatric social worker (Levitt & Goldschmied 1990) carries out the principles of therapy and management, with emphasis on physiotherapy and occupational therapy.

Summary

This chapter provides basic information for planning treatment and management.

(1) The child should be seen as having primarily a motor impairment but may have individual associated impairments due to the brain damage. The motor and other functional disabilities are created by some of the impairments as well as by lack of many everyday learning experiences in various environments.
(2) There is an interaction between the communication, intellectual, sensory, perceptual and motor functions. Physiotherapists therefore consider the influence of associated disabilities on the motor programmes.
(3) Treatment is aimed at impairments and developing gross-motor and fine-motor functions which involve procedures for individual combinations of:
   - Postural mechanisms of balance
   - Movement patterns (synergies) of voluntary movement, including hand function
   - Strengthening for weakness of various kinds
   - Minimising hypertonicity, hypotonicity and involuntary movements
   - Improving postural alignments and patterns of gait
   - Improving ranges of motion of muscles, joints and soft tissues.

Views differ on the significance of specific problems and also on the relationship between them. Therapists need to make careful assessments to clarify the impairment and functional problems of an individual with cerebral palsy, and reflect on their relationships.

(4) Therapy programmes should not have a strict adherence to specific diagnostic classifications, and aetiology may not always influence the treatment used by therapists.

(5) Emphasis needs to be given to the daily functional activities and a participation in life situations, which are priorities of a child or an adult with disabilities and of their families.

(6) The various impairments are preferably treated *in the context of* total daily functions as well as in specific treatments for individuals. The emphasis is therefore less on isolated treatments and more on integrating therapy of impairments within developmental functional training.

(7) The therapist always needs to recognise the emerging functional *abilities and whole functions* within each child's developmental pattern. Normal developmental schedules are only guides and need to be carefully adapted.

(8) Management and therapy are planned from infancy throughout an individual's lifespan to take account of clinical change and different circumstances in an individual's home, schools and community. Management focuses on educating all those primarily involved with a person with cerebral palsy. Chapter 2 discusses this in more detail in a collaborative learning approach. Management also involves working with any other disciplines involved with an individual with cerebral palsy.

(9) Physiotherapists and occupational therapists need to integrate motor learning principles in their therapy programmes. Motor learning models need to encompass emotional, cultural and social issues.

(10) Treatment and management need to commence as early as possible for parental support, parent–child relationships and to promote a child's motor activity as well as minimise any musculoskeletal problems.

(11) The model suggested by the ICF is a general guide to assessment and planning of therapy and management, which matches much that is discussed in this book.

(12) Promoting positive motor experience is key for motivating the best therapy.