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The clinical picture for therapy and management

Cerebral palsy, the most common cause of childhood disability, is the name for 'a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing foetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication and behaviour, by epilepsy and by secondary musculoskeletal problems' (Rosenbaum et al. 2007a). Parents, families, and children are likely to experience emotional and social difficulties. The severity of the cerebral palsy itself may range from total dependency, extremely limited communication, and immobility to the ability to talk, carry out independent self-care tasks, walk, and run. Mild to severe motor difficulties are usually experienced by children according to their severity. Many children and young people with a diagnosis of cerebral palsy are able to access mainstream primary, secondary, and further education. Changes to legislation, advances in technology, and increasing positive attitudes towards disability in society have resulted in opportunities for individuals with cerebral palsy.

Incidence of cerebral palsy

The prevalence of cerebral palsy has consistently been reported to be about 2–2.5 per 1000 live births (1 in every 400 children) over the last 20 years in the Western world. However, in some cases, motor delay that initially presents as cerebral palsy may not be 'cerebral palsy' but a more generalised developmental delay resulting from other conditions such as learning disabilities, progressive neurological conditions, and genetic conditions that can only be recognised later, when a definite diagnosis can be made (Cans 2000).

Motor dysfunction

The motor dysfunction results in disorganised and delayed development of the neurological mechanisms of postural control, balance, and movement. The muscles activated for these motor aspects are

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therefore inefficient and uncoordinated. Individuals have specific impairments of tone, weakness, atypical patterns of muscle activation, and excessive co-contractions. There are absent or poor isolated movements (poor selective motor control), atypical postures, and problems with the use of hands in daily tasks. Biomechanical difficulties result from both the neuromuscular dysfunction and secondary musculoskeletal problems, which add to this complex picture.

The motor dysfunction changes with both growth and the individual's whole development. Change particularly depends on how individuals use their bodies. Although the brain damage is not progressive, the motor and musculoskeletal problems may increase in late childhood, adolescence, and adulthood; physiotherapy and occupational therapy is needed to prevent or minimise these.

What matters most to a child, parents, and family is the child's difficulties in function and participation in daily life. Therapists need to address these difficulties together with a child and parents, or directly with an adolescent and older person with cerebral palsy (see Chapters 2, 3, and 4). Therapists will assess the motor abilities of an individual and detect emerging functional abilities that can contribute to functional activities. It is also encouraging to know that functional activities and participation in daily life can be developed and difficulties minimised even though basic impairments cannot strictly be cured.

Professionals also differ in their views as to which impairments can be minimised, and, if not, when to compensate for these, such as by using task adaptation or equipment to promote engagement in functional tasks. Views or opinions on the underlying motor dyscontrol can be controversial, which is not surprising given that not all the neurological mechanisms are fully understood. Research into the basic dyscontrol and its related biomechanics is continuing.

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 has been further developed and continued with current contributions in the new edition of this book. As many colleagues are now not wedded to any one system of therapy, selections of their views are presented as well as those from our own studies and experience. Current research studies are also given as evidence for specific clinical techniques that need to be part of the eclectic approach.

As a child does not 'move by neurophysiology alone' various ideas of motor and other learning have been integrated into the general therapy framework. The influence of the environmental and social contexts of a child's function are considered as we know that learning takes place in a child's home, school, and community. A child learns best in a familiar environment when therapy is part of a child's activities that are meaningful to that child. It is primarily a child's own intrinsic motivation, encouraged and supported by people in a child's daily life as well as by therapists, that has a profound impact on the child's achievements. Adolescents and older individuals also need consideration of their specific contexts. Many individuals with cerebral palsy face physical indoor and outdoor environmental constraints, differing social attitudes, and sometimes poor availability of services, all of which can be a barrier to their functional abilities. Therapists, together with individuals and their parents, share their strategies to overcome barriers and provide facilitators throughout an individual's life.

Associated impairments and disabilities

Kilincaslan and Mukaddes (2009), in a large clinical trial, suggested that children with a diagnosis of cerebral palsy are also at risk for other neurodevelopmental disorders such as autism, which adds to parental concerns. Brain damage in cerebral palsy may also be responsible for severe cortical visual impairment (Dutton and Bax 2010; Dufresne et al. 2014) and hearing impairments. Many children are also found to have visual problems such as squint (strabismus) and short or long sight (Odding et al. 2006; Dufresne et al. 2014; Dutton et al. 2014). Hearing impairment is not as common (Odding et al. 2006). Children may have problems with non-verbal communication, speech, and language, as well as experiencing aberrations of perception. It is thought that visual perceptual difficulties may be experienced by around 50% of children with cerebral palsy (Ego et al. 2015). However, self-directed movement through space leads to the development of visual–spatial competencies (Campos et al. 2000; Coté 2015), which suggests that children with cerebral palsy are likely to develop these skills more slowly.

A number of children with cerebral palsy have difficulties performing certain movements, even though there is no paralysis, because the patterns have not developed. This is sometimes described using the umbrella term of *dyspraxia* (Lee 2004). This can involve movements of limbs, face, eyes, or tongue. There may be disorders of memory of motor sequences (Lesný et al. 1990). Tactile sensation, proprioception, two-point discrimination, and perception may be impaired (Odding et al. 2006). A study by Yekutiel et al. (1994) found sensory impairment in the hands of children, and more recent studies suggest that tactile deficits are prevalent in over 70% of children with unilateral cerebral palsy (Auld et al. 2012). Some children may also have various behavioural problems such as distractibility and hyperkinesis, which are linked to the brain damage. Emotional relationships and social factors may also give rise to behaviour difficulties. Specific behaviour problems and psychological difficulties are associated with epilepsy (Carlsson et al. 2008). Learning difficulties and various epilepsies (seizures) at birth, or appearing in later childhood, may be present (Himmelmann et al. 2006; Odding et al. 2006).

Not every child has some or all of these associated impairments. In some cases the associated impairments or disabilities may present greater problems than the motor conditions. Severity of epilepsy, cognitive difficulties, communication problems, and excessive behaviour problems are such examples. Some epilepsies can be controlled with medication, but others may be more challenging and severe.

Even if the motor impairments were the only concern, the resulting paucity of movement would prevent the child from fully exploring the environment. Therefore acquisition of sensations and perceptions of everyday things are limited. 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 may slow down the development of speech and language. General understanding may suffer, so a child appears to have an intellectual disability. This can go so far that good intelligence has been camouflaged by severe physical disability. Cognitive assessment is therefore crucial if we are to ensure that we neither under- nor overestimate a child's intelligence. Furthermore, the problems of movement can affect the general behaviour of a child due to the lack of satisfying emotional and social experiences for which

movement is necessary. However, positive attitudes in a family and a child's personality can encourage optimum emotional and social development. They, together with their therapists, will find experiences for the child's whole development.

The population of people with cerebral palsy is heterogeneous. Different individual impairments and dysfunctions in, say, cognition, language, or movement have the potential to contribute to reduced participation in daily life. The correlation between abilities and participation is therefore extremely individual and cannot be presumed.

Teamwork. In order to manage the multiple disabilities and lack of related learning experiences that interfere with a child's development, a physiotherapist or occupational therapist needs to be part of a team. Teams of different specialised professionals contribute programmes for impairments, motor functions, and daily tasks to increase participation by children and their families in daily life situations. Teamwork is discussed in Chapters 2, 3, 4, 8, 11, and 12.

Broad framework for therapy and management

The World Health Organization's (WHO) current International Classification of Functioning, Disability and Health (ICF) (WHO 2001, 2013) describes a person's functioning in terms body functions, body structures, activities, participation, environmental factors, and personal factors, which are defined below.

These components may all interact with each other but, in a particular situation, only a few interactions are likely to be important. The therapist needs to explore the components for an individual and consider if, and how, they interact.

Knowledgeable therapists have been taking account of all the ICF components for many years, although they often used different terminology in the past. The advantage of using the ICF model is that it provides a common language for use in different clinical disciplines, while also bringing clinicians together. Therapists need to focus more on the functional outcomes rather than only on reducing underlying impairments (Imms et al. 2016).

The ICF framework is relevant to many existing procedures in clinical practice, including assessment, goal setting, evaluating outcomes, and communicating with colleagues and with the person and everyone involved. The use of the ICF can encourage a more holistic and comprehensive assessment and management of people with health conditions.

Definitions used in the ICF

Functioning an umbrella term for body functions, body structures, activities, and participation. It denotes the positive aspects of the interaction between an individual and that individual's environmental and personal factors.

Body functions physiological functions of body systems.

Body structures anatomical parts of the body and limbs.

Activity the execution of a task or action by an individual such as walking or using hands. This includes daily living tasks such as eating and toileting. However, these tasks can also be participation in life situations.

Participation involvement in everyday life situations such as interactions with family members or friends. Participating at school and in a community is promoted. Activities and participation are often linked.

Disability an umbrella term for impairments, activity limitations, and participation restrictions. It denotes the negative aspects of the interaction between an individual and that individual's environmental and personal factors.

Impairments problems in body function or structure.

Activity limitations difficulties an individual may have in executing activities.

Participation restrictions problems an individual may experience in involvement in life situations.

Contextual factors

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

Personal factors influence how disability is experienced by an individual. These include age, coping styles, and drive. Personal factors are not currently classified in the ICF as they can vary widely between different cultures.

The following examples are influenced by Schiariti et al. (2015), who derived core sets of the ICF for children and youth with cerebral palsy, which are shortlists of ICF categories that are considered most relevant. However, it should be noted that the introduction of a version of the ICF for children and young people has since been combined into the main ICF and therefore no longer exists as a separate framework.

Examples in cerebral palsy

Body functions

Physiology, neurophysiology, respiratory, genitourinary, vision, and other sensory systems (including proprioception). Sensation of pain. Fitness and strength. Muscle tone. Sleep. Range of motion. Control of balance, voluntary movement, and coordination.

Body structures

Anatomy. Musculoskeletal changes due to natural history and other factors.

Activity

Maintaining and changing head control, lying, sitting, squatting, kneeling, and standing. Mobility such as rolling, crawling, bottom-shuffling, walking forwards, backwards, and sideways (including cruising), running. Speed, distance, and endurance are considered. Arm and fine hand functions such as reach and grasp, pick up, release, manipulate. Achievement of eating, drinking, toileting, washing, playing. Therefore, development of activities both basic and complex in childhood are planned.

Participation

Involvement in the individual's community, taking part in school, sports, shopping, use of playgrounds, and social events. Relationships with parents, family members, and relatives. Using equipment, technology, and specialist equipment. Coping with transport. Learning to eat in a culturally appropriate way as well as culturally appropriate physical contact.

Environmental factors

These are external to an individual. Architectural barriers to access, terrain, and climate. Use of mobility aids in specific environments both indoor and outdoor. Family and social attitudes. Availability of health and rehabilitation services and of assistive equipment and technology.

Personal factors

Age, sex, personality, coping styles, drive, culture, character, overall behaviour, and self-perception or self-esteem. Past and current life experiences.

While quality of life and health-related quality of life are not directly included in the ICF, many researchers have demonstrated the link between increased participation, health, and well-being (Anaby et al. 2017). Bjornson et al. (2008), in their

research with young people aged 10 and over, emphasised participation. However, they found that 'functional level and performance did not influence quality of life'. Livingston et al. (2007) emphasise that quality of life is very subjective, as recognised by Albrecht and Devlieger (1999).

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 specialist equipment such as powered wheelchairs, computers, and other technology are provided. Depending on the severity of cerebral palsy, innovative functional strategies or motor compensations may allow independent function with little focus on impairments. There are a number of examples, such as the MOVE programme (Bidabe and Lollar 1990; Thomson 2005). There is also an independent functional programme within conductive education (Hari and Akos 1988). Many research studies in constraint-induced movement therapy (Charles et al. 2006; Wallen et al. 2011) and in hand–arm bimanual intensive therapy (Charles and Gordon 2006) showed that they did not change impairment but improved function in the arm and hand of children with hemiplegia and coordination between both upper limbs. However, Bjornson et al. (2013) in their research found that achievement of *capacity* in a structured clinical setting did not transfer to *participation* in everyday life.

There are three points supported by clinical experience:

1. owing to the brain damage, not all impairments can be minimised
2. when selected impairments have been minimised, this did not always carry over into daily activities
3. daily activities are especially linked with participation.

However, secondary impairments such as contractures and musculoskeletal pain may result from primary motor difficulties. Impairments such as specific weakness, poor balance, atypical coordination, and hypertonus may increase with time. These secondary impairments may limit independence of some activities and restrict participation in an individual's home and community over time.

The overall goal of those working with children and young people with cerebral palsy or motor delay is therefore understood to be increased participation in life situations. However, as individual therapists with different professional backgrounds we will work with the child towards this goal in different ways, and it is important that we are able to explain and promote the uniqueness of our professional role in order to demonstrate the value we bring to the multidisciplinary team.

The goals of therapy are to develop:

1. forms of communication (gesture, speech, and the use of alternative forms of communication with pictures, signs, or electronic aids)
2. independence in the daily activities of eating, drinking, dressing, washing, toileting, and general self-care with and without aids, such as adapted utensils, bath aids, and other specialist equipment
3. abilities to play and achieve hobbies and recreational activities with or without adapted toys and equipment
4. some form of locomotion and independent mobility, which may include walking aids, wheelchairs, pedal karts, tricycles; or adapted motor vehicles; powered wheelchairs and powered mobility equipment for very young children, usually from age 2 years or even 11–12 months, if there are no comorbidities.

All these goals need to be considered in terms of learning processes interacting with

neurological and orthopaedic aspects and environmental constraints. Communication between therapists and people with cerebral palsy is fundamental in therapy and learning processes. 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. Learning models need to give adequate attention to this fact. Consideration of cultural factors in planning the programme is very important.

Aetiology of cerebral palsy

Premature infants, very low birth weight, multiple births, and intrauterine infection present a greater risk for cerebral palsy. There are many causal pathways rather than one cause for the brain damage. These include abnormal development of the brain, anoxia, intracranial bleeding, excessive neonatal asphyxia (hypoxic ischaemic neonatal encephalopathy), perinatal stroke, hypoglycaemia, or neurotrophic virus. The complexity of causes has been extensively discussed in the medical literature (Himmelmann et al. 2010; MacLennan et al. 2015; Hagberg et al. 2016).

Genetic causes

There are few known genetic causes for cerebral palsy (Rosenbaum and Rosenbloom 2012: chapter 3). Schaefer (2008) mentions genetic pathways that may predispose to cerebral palsy. However, there are other genetic progressive disorders that may seem to be cerebral palsy; for example, Lesch–Nyhan syndrome with symmetrical spastic paraplegia or dystonia, with or without dyskinesia, or Rett syndrome, which has some features seen in cerebral palsy. Global developmental delay or learning disabilities may result from a genetic cause and, as described above, the early motor delay may appear to be cerebral palsy. The Surveillance of Cerebral Palsy in Europe collaboration (Cans 2000) has a recommendation that a cerebral palsy diagnosis should be confirmed by the age of 4 years.

Causes in later childhood

Although the definition of cerebral palsy gives the impairment occurring in the fetal or infant brain, usually agreed to be up to 2 years, there are causes of brain damage in later childhood such as trauma, road accidents, anoxia as in near-drowning, choking, and from various infections, such as meningitis and cerebral malaria. The subsequent brain injuries share common features with cerebral palsy and as such require similar assessments and interventions as well as the provision of additional support for parents and families.

Therapists are not always guided by the aetiology in 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 be acquainted 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 of problems during neonatal or prematurity periods may sometimes give an indication of the causes of cerebral palsy. Early neuroimaging detects lesions likely to result in cerebral palsy.

Neuroimaging

The continuing advances in neuroimaging such as magnetic resonance imaging (MRI)

and computed tomography (CT) help in identifying the extent and site of the neurological lesions, with neuroimaging being abnormal in more than 80% of children with cerebral palsy (Himmelmann et al. 2017). There are variations in brain lesions due to the timing of the insult that relate to clinical motor severity and associated impairments (Krägeloh-Mann and Horber 2007; Korzeniewski et al. 2008). Brain scans locate lesions in the white matter that are described as periventricular leukomalacia. These lesions may be associated with spastic diplegia, a diagnosis that has a wide spectrum of disability. However, spastic diplegia (bilateral cerebral palsy with lower limbs more severely affected) is also related to other causes (Rosenbaum and Rosenbloom 2012: chapter 3). Although the diagnosing of cerebral palsy remains ultimately dependent on the history, regular observations by both parents and clinicians and careful clinical examinations by those with experience of working with children with cerebral palsy, the Surveillance of Cerebral Palsy in Europe collaboration has suggested the introduction of an MRI classification system that might support better understanding of the relationship between brain structure and functional abilities.

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. Therapists must therefore not think of themselves as treating an upper motor neurone lesion in a 'little adult', nor can they regard the problem solely as one of delayed development. What the therapist faces is a complex situation of pathological symptoms within the context of a developing child (Illingworth et al. 2012; Sharma and Cockerill 2014 (an updated version of Mary Sheridan's work)).

There are six main aspects to the clinical picture, as follows.

1. Delay in the development of new skills expected at a child's chronological age.
2. Persistence of immature behaviours such as feeding difficulties and retained primitive reflexes that sometimes interfere with function.
3. Slow rate of progress from one developmental stage to the next.
4. A smaller variety of skills in comparison with a child who is developing typically.
5. Marked variations in the sequence of skills greater than that which we might see in typical development.
6. Atypical and unusual performance of skills.

In order to recognise any atypical motor patterns, therapists should know what a typical child does and understand the 'expected' patterns of behaviour at the various stages of child development. Reference will have to be made to the extensive literature on the field of child development. Typically developing children show variations from the 'normal' sequences and patterns of development that have been derived from the *average* child. Parents or family members may themselves identify differences in their child's development and want referral to professionals to discuss their concerns. Professional specialists in cerebral palsy will assess whether differences are in fact normal variations. Information on each individual child's developmental abilities and difficulties should be sought from the consultants, therapists, and others in the team.

Other variations need not necessarily convey a diagnosis of cerebral palsy.

Both the climate and cultural differences affect typical motor development (Solomons and Solomons 1975; Hopkins and Westra 1989; Leavitt 1999). However, in any culture, children with cerebral palsy will show additional variations due to neurological and biomechanical causes.

If one considers, say, the developmental scales of gross motor development, in cerebral palsy a child has frequently achieved abilities in motor functions at one level of development, omitted abilities at another level, and only partially achieved motor abilities and functional components at still other levels. There is thus often more of a scatter of abilities as well as whole motor functions than found in typical children. (Details of development are discussed in Chapters 7, 9, and 10.)

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

Although typical child development is the basis on which the atypical development is appreciated, it does not follow that assessment and treatment should rely upon a strict adherence to typical developmental schedules. Therefore, the developmental schedules in typical child development should only be used as *guidelines* in treatment, and adaptation should be made for each child's development, abilities, and individuality.

More attention is usually given to motor development than other avenues of development, as it is the motor dysfunction that characterises cerebral palsy. Here again, the therapist should remember that 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 having its own pattern or avenue of development. Furthermore, the potential for function is dependent not only on the abilities and disabilities present but also on a child's personality and 'drive' as well as a capacity to learn. Therefore, a total habilitation programme is necessary and should be planned to address the whole development of each child, adolescent, and adult.

While aiming to maximise functional abilities, the therapists concerned must take account of the *damaged* nervous system and adjust their expectations of achievements by individual children. Cerebral palsy cannot be cured. However, children will continue to develop to reach their own potential. The reliability of prognoses depends on a therapist's clinical experience as multiple factors are usually involved. Prognosis is discussed in Chapters 8 and 9.

Change at different ages

As the lesion is in a developing nervous system, the clinical picture is clearly not static. As more is demanded of the child, the degree of the disability appears to be greater. For example, a 3-year-old is expected to do more than a baby, and therefore the difficulties are greater for the same pathology.

In addition, the pathological symptoms may develop over time. Babies may often be hypotonic, becoming hypertonic later. Involuntary movements or dystonia 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. Cerebral palsies have an emerging diagnosis. Later, especially in adolescence, growth and increase in weight may contribute

to apparent deterioration. Recent research identifies that deterioration is not inevitable in all cases (see Chapters 4 and 9). Changes in function and health are also evident in adolescents and adults.

Treatment and management in infancy until age 2–3 years. The earlier the treatment is started, the more opportunity is given for whatever potential there may be for developing any abilities and for decreasing the movement and postural difficulties (Spittle et al. 2015; Morgan et al. 2016a), although at present there is limited empirical evidence regarding the effect of early intervention on motor skill development (Hadders-Algra et al. 2016). There has been an increase in specialised neonatal therapists (physiotherapists, occupational therapists, and speech and language therapists (SaLTs)) who have developed evidence-based assessments and therapy (see Chapter 8). Guidelines produced by the National Institute for Health and Care Excellence (NICE 2017) suggest means of recognising the early motor features of cerebral palsy and call for all children who are at risk of developing cerebral palsy to be referred for an urgent multidisciplinary assessment. A systematic review of the literature (Novak et al. 2013) also calls for a move away from 'conservative late diagnosis' in order that early intervention can be utilised to optimise neuroplasticity.

Therapists offer a variety of pleasurable and developmentally appropriate activities, including motor activities, enjoyed by both parent and baby. During intervention, therapists observe if babies or young children make their own efforts to move using compensatory or adaptive patterns that 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 later deformities. Early therapeutic input minimises such problems.

However, abnormalities detected in premature and term infants may be transient as some infants overcome them without intervention. Nelson and Ellenberg (1982), studying a large sample of infants suspected of cerebral palsy, found that many with marked neurological signs may later prove to be only mildly affected or go on to develop typically. It is possible to make a 'false-positive' early diagnosis (Bosanquet et al. 2013). De Groot (2000) made an intensive study of posture and motility in preterm infants. There are assessments of preterm, term, and young infants that predict cerebral palsy if used by therapists trained in these methods (Lacey et al. 2004; Einspieler et al. 2005). Marcroft et al. (2014) found few studies of value predicting cerebral palsy using the Lacey Assessment of Preterm Infants. We still need to know more about which babies are likely to 'come right' on their own or turn out to have a progressive neurological condition rather than cerebral palsy. Nevertheless it is better to provide them with the benefits of developmental therapy. Blauw-Hospers and Hadders-Algra (2005) 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 value of early developmental intervention is to provide an increase in babies' sensory–motor and everyday experiences and especially develop interaction with their parent(s). However, there is a need to minimise sensory stimulation and handling if an infant is very preterm. The sooner a baby can be helped to move, the sooner there is exploration and experience for that baby. The therapist is in fact contributing to learning and understanding as well as enabling babies to bond with their parents or carers. The parents and family are assisted to cope with early rearing of an

infant who is developing atypically. There is clearly no doubt about its importance to the parents, who receive a great deal of practical advice and support from therapists. Parents need practical ideas for feeding, childcare, and playful activities for their child throughout any emerging diagnoses.

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 as well as the individual's community. People 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 the lifespan of each person with cerebral palsy.* Management will include working with other allied health professionals such as SaLTs and orthotists, as well as orthopaedic surgeons and other medical consultants. Therapists will share selected skills and advice on equipment with a child's parents and anybody closely involved with any person with cerebral palsy and motor delay. Extra therapy sessions by professionals will not achieve as much as combining treatment with working with parents and family. Therapists need to carry out a detailed assessment of a child's function, identifying any motor impairments that are acting as a barrier to each activity. Consideration of a child's or young person's functional abilities and difficulties within their participation in their daily life is essential. Parents and teachers and others involved who know a child or youth will need to be consulted for their valuable observations. Therapists can then work out treatment programmes together with individuals and their family members.

Classification systems

The classifications describe and simplify the levels of function in cerebral palsy, and enable a quality of care and can aid selection of assessment tools. Their use of everyday language enables parents to participate in the development and use of the classifications. Each of the classification systems uses five levels (Paulson and Vargus-Adams 2017).

Gross Motor Function Classification System (GMFCS)

The complete Gross Motor Function Classification System (Palisano et al. 1997, 2008) can be downloaded from the CanChild website (www.canchild.ca).

This describes the level of functional ability, in a developmental picture, using age-dependent criteria. The descriptive features are reported at each level, saying what a child can do in self-initiated motor functions such as sitting, changing postures, crawling, standing, and walking. The level of function decreases from level I to V, with level V representing children with severe motor restrictions. The GMFCS level that is assessed is the performance that a person typically does in home, school, or community.

The classification system can be used to compare individual children of the same age with each other, as they can be functioning at different levels. All children with cerebral palsy are classified according to what they can do (Gorter et al. 2004). The quality of movement is not an important factor. In several centres, doctors and therapists, as well as parents, are consistent in their identification of the level of a child (Morris et al. 2006).

Classification of an individual child gives a shorthand description of a person with cerebral palsy and is particularly useful for communication between professionals and for generally relating therapy suggestions

to a level of ability and disability of an individual. There are increasing levels of function for ages 1–2 years (before second birthday); 2–4 years (between second and fourth birthdays); 4–6 years (between fourth and sixth birthdays); and 6–12 years (between sixth and twelfth birthdays). The classification system was expanded and revised in 2008 for ages 12–18 years (between twelfth and eighteenth birthdays) (Palisano et al. 2008).

The level assigned to a child is not 'set in stone', particularly if it is assigned before the second birthday. Wood and Rosenbaum (2000) carried out a historical study of children using the GMFCS. They followed changes in the GMFCS level from age 1–2 years to age 6–12 years. The data (their table IV) show that only about 40% of 78 children stayed in the same level, with about 20% improving and about 40% getting worse. Two children went up three levels and one child dropped three levels. So the figures show that, although the GMFCS levels of a *group* of children do not change much as they get older, the level of an *individual* child may change dramatically.

Gorter et al. (2009) found that only about 60% of children classified at age 18 months remained in the same level a year later. Therefore, all children should be reclassified after their second birthday.

Reclassifying at a much later age may also need to be considered, as intervention or ageing can have an effect. A study by McCormick et al. (2007) of the stability of the GMFCS in 103 adults showed that about 70% did not change level after age 12 (see their table IV). However, about 10% went up one level and about 20% went down one level.

Example of GMFCS levels

The following list relates to children between their sixth and twelfth birthdays.

Level I Walks at home, school, outdoors, and in the community; climbs stairs without using a railing. Performs gross motor skills such as running and jumping with limitations of speed, balance, and coordination. May participate in sports.

Level II Walks in most settings; difficulties in walking when carrying objects. Uses physical assistance, hand-held walking devices when there are difficulties with long distances, inclines, uneven terrain, confined space, and crowds. Climbs stairs using a railing. Children have at best only minimal performance of skills such as running and jumping. Adaptations needed for sports.

Level III Walks with hand-held mobility devices in most indoor settings; limitations in walking outdoors. Climbs stairs using a railing with supervision or assistance. Use of wheeled mobility over long distances and depending on arm function may self-propel a manual wheelchair for sports.

Level IV Walks short distances with physical assistance or uses powered mobility. When positioned, may use a body-support walker. At school and in the community children are transported in a manual wheelchair or use power mobility. Adapted seating is required. Independent floor mobility may be used at home. Physical assistance or powered mobility needed for sports.

Level V Transported in a manual wheelchair in all settings. Some achieve self-mobility using powered mobility with extensive adaptation of seating. Children have limited control of head, trunk, arms, and legs.

Manual Ability Classification System (MACS)

The complete Manual Ability Classification System for children with cerebral palsy 4–18 years (Eliasson et al. 2006, 2010) can

be downloaded from the MACS website (www.macs.nu).

This classification describes children's ability in self-initiated hand functions at home, school, or community. The MACS and GMFCS classifications do not give the reason for the levels of ability.

This is a valid and reliable way for therapists to classify hand function (Eliasson et al. 2006, 2010; McConnell et al. 2011; Öhrvall et al. 2014).

Level I Handles objects easily and successfully.
Level II Handles most objects but with somewhat reduced quality and/or speed.
Level III Handles objects with difficulty; needs help to prepare and/or modify activities.
Level IV Handles a limited selection of easily managed objects in adapted situations.
Level V Does not handle objects and has severely limited ability to perform even simple actions.

Eliasson et al. (2006) compared the GMFCS and MACS levels of 168 children and concluded that the GMFCS level agreed with that of the MACS in only around half of the children.

MACS is extended through the Mini-MACS, which is designed for children between the ages of 1 and 4 years; early studies of the Mini-MACS have demonstrated good inter-rater reliability (Eliasson et al. 2017) between parents and therapists and between therapists. It is necessary to ask a parent or someone who knows each child well rather than use a specific test.

In the Mini-MACS, levels I, II, and V are identical to the MACS but level III is just 'handles objects with difficulty' and level IV has 'simple actions' instead of 'adapted situations'.

Bimanual Fine Motor Function (BFMF) classification

The Bimanual Fine Motor Function classification (Beckung and Hagberg 2002; revised and validated by Elvrum et al. 2016) classifies fine motor function in children with cerebral palsy aged 3–18 years and has its own five levels.

Beckung and Hagberg (2002) compared the GMFCS and BFMF levels of 176 children and concluded that the GMFCS level agreed with that of the BFMF in just over half of the children. However, nearly 30% of the children were classified in level I for both GMFCS and BFMF.

Carnahan et al. (2007) present a retrospective study of 365 children with cerebral palsy, also showing that gross motor function and manual ability are often discrepant in children with cerebral palsy and that the patterns seem to vary across the different topographical classifications (see section 'Topographical classifications').

Communication Function Classification System (CFCS)

The complete Communication Function Classification System for individuals with cerebral palsy (Hidecker et al. 2011) can be downloaded from the CFCS website (www.cfcs.us). This classifies levels of everyday communication effectiveness.

Level I Sends and receives with familiar and unfamiliar partners effectively and efficiently.
Level II Sends and receives with familiar and unfamiliar partners but may need extra time.
Level III Sends and receives with familiar partners effectively, but not with unfamiliar partners.
Level IV Inconsistently sends and/or receives even with familiar partners.
Level V Seldom effectively sends and receives, even with familiar partners.

All methods of communication are considered such as speech, gestures, behaviours, eye gaze, facial expressions, and augmentative and alternative communication (AAC).

AAC includes manual sign pictures, communication boards, and talking devices (voice output communication aids or speech-generating devices).

Helen Cockerill (2011), a specialist speech and language paediatric therapist, has commented on the CFCS.

Eating and Drinking Ability Classification System (EDACS)

The Eating and Drinking Ability Classification System (Sellers et al. 2014) classifies the safety and efficiency of eating and drinking from age 3 years onwards. The usual performance is observed. Sellers et al. (2014) developed this classification, stating its reliability.

SaLTs have confirmed its reliability. This does not replace a comprehensive professional assessment by an individual's SaLT and may include videofluoroscopy assessment of dysphagia and oromotor measures.

Level I Eats and drinks independently with safety and efficiency with no difference from peers. May cough or gag with challenging food textures but manages a large range of textures.

Level II Eats and drinks safely with some food loss and more time needed to complete a meal than peers. Manages a large range of textures but coughs/gags on challenging food textures.

Level III Eats and drinks with some limitations to safety and efficiency. Coughs with fast flowing liquids or large food quantity and challenging food textures.

Level IV Significant safety risks, cannot swallow drink or food without risks of aspiration. Oral feeding is possible with mashed or pureed food.

Level V High risk of aspiration with feeding and drinking. Selected tube-feeding methods. Manages small tastes or flavours.

Efficiency refers to the length of time and effort required to eat or drink, as well as whether food or drink is kept in the mouth without loss. This avoids tiring during a meal.

Safety risks include choking when a piece of food becomes lodged in the airway as well as aspiration when food or fluid enters the lungs.

Levels of assistance required

1. Independent.
2. Requires assistance.
3. Totally dependent.

There is a review and commentary on EDACS by Sophie Scott (2014) in terms of risk and functional ability. She includes sucking, biting, chewing, swallowing, and keeping food and fluids in the mouth. Mention is made of different settings, skill, and the quality of the relationship with familiar/unfamiliar carers.

Association between the functional classifications

Compagnone et al. (2014) demonstrated a positive correlation between the GMFCS, MACS, and CFCS levels in 87 children, although the most significant association was specifically in the children in level V.

Topographical classifications

Bilateral cerebral palsy (affecting both sides of the body)

Involvement of all limbs and body. Arms are equally or more affected than the legs. Many are asymmetrical. *(Quadriplegia (tetraplegia) or triplegia is used in some research.)*

Involvement of limbs, with arms much less affected than legs and body. Asymmetry may be present. *(Diplegia is used in some research and by some clinicians.)*

Unilateral cerebral palsy

Limbs and body on one side are affected. (*Hemiplegia is used in some research and by some clinicians (Neville and Goodman 2001).*)

Gorter et al. (2004) found that hemiplegia was mainly in GMFCS level I, but others were in levels II–IV. Diplegia was found in all levels. Quadriplegia was mainly in levels IV and V but Gorter et al. found a substantial proportion were in levels I and II.

These topographical classifications can be imprecise, as they may change with a child's development. Active use of one upper limb together with both lower limbs or use of two upper limbs with one lower limb may convey a triplegia that could become quadriplegia. Upper limbs may appear unaffected, suggesting paraplegia, but being really a diplegia with only fine hand use being affected when this is later expected. There may be increasing disability in hand/arm function so that there is closeness from diplegia to quadriplegia. Individuals with hemiplegia may experience minor involvement on the unaffected side. Perceptual or motor planning difficulties may be found on the unaffected side. A monoplegia is rare, usually becoming a hemiplegia with increased active motor development. It is for these reasons that many clinicians are moving towards functional classifications that are based on a child's ability to engage in activities rather than on their specific physical impairments; this is discussed later in the chapter.

Additional descriptors of the motor type experienced by the child provide the clinician with additional information, for example *bilateral dyskinetic cerebral palsy* or *unilateral spastic cerebral palsy.* Comments on the presence of any asymmetry also help clinicians to share information about the child.

Classification of motor types of cerebral palsies

The predominant impairments will contribute to the diagnostic type referred for therapy and management. There are also mixed types of cerebral palsy where the characteristics of more than one type are present.

Spastic cerebral palsy

The main motor characteristics are as follows.

Spasticity. If spastic muscles are stretched at a particular speed, they respond in an exaggerated fashion. There is a movement block called a 'catch'. Spasticity is a velocity-dependent resistance of a muscle to stretch, and resistance differs with the direction of joint movement, for example flexion versus extension (Sanger et al. 2003). Second, resistance to externally imposed movement rises rapidly above a threshold speed or joint angle after the 'catch' is felt (Sanger et al. 2003).

There are increased tendon jerks, occasional clonus, and other signs of upper motor neurone lesion. Stiffness is not true spasticity and may accompany the hyperactive reaction to brisk passive stretch. Muscle and soft tissue changes are causes of stiffness with inertia of the limbs (Katz and Rymer 1989; Dietz and Berger 1995).

There are other important aspects that need consideration, such as the recognition of incoordinated movements, weakness and inefficient muscle work, as well as compensations for absent or atypical postural mechanisms. Later there is an increase in growth of the bones compared with short muscles failing to keep up.

Shortening hypertonic ('hypoextensible') muscles pull the joints into atypical

motor patterns involving the whole child or at least of the whole limb. Shortland et al. (2002) and Fry et al. (2004) have used ultrasound to study muscles in spastic cerebral palsy, and one finding was that weakness of muscles contributes to muscle shortening and atrophy. At first 'hypoextensible' muscles with or without hypertonic stiffness can be overcome in a young child, but later they can become fixed. In time, atypical postures and movements that are unfixed (dynamic) deformities may become fixed deformities (contractures) with shortening of ligaments, connective tissues, and tendons and decreasing joint ranges Subsequently this can lead to hip dislocations, scoliosis, and bony deformities. Atypical postures, deformities, and contractures, particularly in the upright positions, contribute to atypical gaits. Atypical gaits are described in more detail in Chapter 12 and gait analysis is described in Chapter 8.

Some clinicians may still use 'spasticity' and 'spastic muscles' as an umbrella term for stiffness of limbs, but it is important that the difference between physiological spasticity and stiffness is understood in order that the most effective treatment interventions can be offered from both a therapeutic and medical perspective. The hyperactive stretch reflex is *not* the cause of dysfunction but the coexisting motor disorders, particularly weakness, are significant (Lin 2004, 2011; Ross and Engsberg 2007). Movements are usually slower than the velocity needed to obtain the hyperactive stretch reflex.

Postural alignments (body shape) (see Figs 1.1–1.3*).* These are often extensors in the leg and flexors in the arm. However, the therapist will find *many* variations on this, especially when the child reaches different stages of development. The atypical limb postures become held by stiffness with shorter 'spastic hypertonic' muscles whose lengthened antagonists are weak, in that they cannot overcome the tight pull of the

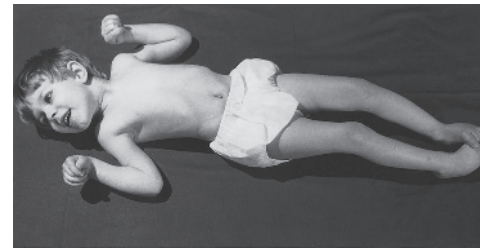

Figure 1.1. Child with bilateral spastic quadriplegia. Head preference to right, shoulders protracted, semiabduction, elbows flexed–pronated, wrists and fingers flexed, thumb adducted. Hips and knees flexed, tendency to internal rotation–adduction with feet in equinovarus, toes flexed.

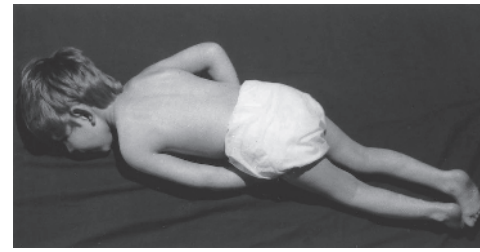

Figure 1.2. Same child with bilateral spastic quadriplegia with postural changes in prone. Asymmetry of arms caught under body. Hips and knees flexed, feet in equinovarus. Head preference is now to left.

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. A symmetrical trunk.

shortened muscles and so correct the atypical postures. The short muscle groups are also weak and cannot easily assume a typical alignment.

Many children have floppiness of the head and trunk together with the stiff spastic hypertonic limbs. This is associated with delayed development of the mechanisms of postural stabilisation and postural adjustment of the head and trunk as well as the pelvic and shoulder girdles. Besides stiffness and weakness of limbs there are atypical compensations of all the postural mechanisms unavailable for balance control. In all editions of this book, the postural mechanisms are and were given more importance so that spasticity is not overemphasised as a cause of dysfunction. Lin (2011) has the same view of spasticity.

Muscle hypertonus is defined as resistance to externally imposed movement about a joint and there are different types of hypertonus. There is spastic hypertonus, rigid hypertonus or dystonic hypertonus, and mixed hypertonus, which are differentiated by Sanger et al. (2003). Spastic hypertonus particularly affects deformity but not motor function, which depends more on weakness. Rigidity is recognised by a continuous resistance to passive stretch throughout the full range of motion. This *lead pipe* rigidity is considered less common in cerebral palsy. If present it can be differentiated from dystonia as well as from spasticity (Lin 2004). Spasticity is not continuous but offers resistance at a point or small part in the speed of the passive range of motion. This is discussed further in section 'Dyskinetic cerebral palsy (dystonic and choreoathetotic)'. Drugs such as botulinum toxin A and oral and intrathecal baclofen are used to control spasticity and dystonia (Albright and Neville 2000; Lin 2004), together with a physiotherapy or occupational therapy programme (Novak et al. 2013). Focus on spasticity includes the use of neurosurgical selective dorsal rhizotomy (Peacock and Staudt 1991; McLaughlin et al. 2002; Tedroff et al. 2011; Josenby et al. 2012; Ailon et al. 2015).

The common atypical postures and movements in supine, prone, sitting, standing, walking, and hand function are described in more detail with therapy and management suggestions in Chapter 9.

Atypical postures and stiffness may increase with excitement, fear, or anxiety as well as when pain is present. Shifts in spastic hypertonus may occur in the same affected parts of the body or from one part of the body to another in, say, stimulation of remnants of reflex activity. Changes in hypertonus are seen with changes of position in some children. Position of the head and neck may affect the distribution of spasticity. Any sudden or fast movements, rather than slow movements, increase stiffness.

Voluntary movement. Spasticity does not necessarily mean paralysis. Voluntary motion is present and 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 muscles have been found to be weak. For example, the removal of spasticity of the gastrocnemius with botulinum toxin A injection reveals weak plantarflexion.

The groups of muscles or *chains* of muscles used in the movement synergies (muscle activation patterns) are different from those used by typical children of the same age. Either the muscles which work in association with each other are stereotyped and are occasionally seen in typical infantile levels of movement or the association of muscles is atypical. 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 function. One example of a typical arm pattern is shoulder flexion–adduction with some external rotation for feeding or combing one's hair. In the case of a child with coexisting motor problems, the arm pattern is usually flexion–adduction with *internal* rotation and *pronation* of the elbow. The ability to fractionate movement is very difficult for a 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 typical reciprocal relaxation persists. However, typical co-contraction is also evident in any typical person attempting a new and difficult skill with hands or legs. Before the postural control develops in typical infants there is a co-contraction response in weight bearing and early stages of walking in children without cerebral palsy. These patterns persist in children with motor problems (Lin 2004). 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 movements. There is a lack of isolated or discrete movements (selective motor control) and fine motor coordination is delayed in younger typical children as well as persisting in the spastic type of cerebral palsy.

Associated impairments

1. Intelligence varies and is usually impaired in bilateral spastic quadriplegia.
2. Sensory loss occasionally occurs in unilateral hemiplegia with a visual field loss and lack of sensation in the hand (Yekutiel et al. 1994; Odding et al. 2006). Sensory dysfunction such as sensory discrimination and sensory integration rather more than sensory loss is present in individuals (Lesný et al. 1993). 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, motor planning, and motor memory 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 bilateral spastic quadriplegia.
6. Growth of limbs in unilateral hemiplegia or severely affected lower limbs in bilateral cerebral palsy can be less than the other limbs.
7. Epilepsies are more common in bilateral spastic quadriplegia and unilateral hemiplegia but minimal in bilateral diplegia (Neville 2000).
8. A congenital suprabulbar palsy is found in some individuals with bilateral quadriplegias (Neville 2000). The Worster-Drought syndrome is a rare form of cerebral palsy with often mild motor problems, but often with severe difficulties with oral–motor skills for feeding, saliva control, and speaking.

Dyskinetic cerebral palsy (dystonic and choreoathetotic)

The main motor characteristics are as follows.

Involuntary movements – dyskinesia (athetosis). These are unusual, purposeless movements that 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 (or 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. Involuntary excessive open mouth smiling often facilitates socialisation. Dyskinesia may appear only in hands or feet or in proximal joints, or in both distal and proximal joints. Generally the child finds great difficulty in being still.

Chorea is random ongoing sequences of involuntary movements or fragments of them. There is an overlap with athetosis termed choreoathetosis (Sanger et al. 2010).

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 their postural mechanisms. A common pattern is to 'run headlong' using momentum as they cannot stand still on one leg or adjust their posture for slower walking. They run before they can walk.

There are also standing postures in late childhood, adolescence, and adulthood that usually involve backward lean with hip extension, knee semiflexion, lordosis, and kyphosis with chin jutting well forwards and arms held forwards. This is another compensation for instability (Fig. 1.4).

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. Manipulation and some simple hand actions may be difficult. Grasp and release have extreme flexion and extension movements that some older children learn to control for finer grasp or use of large keys on a computer.

Figure 1.4. Person with dystonia/dyskinesia in standing or walking positions. In late childhood, adolescence, and adulthood posture is usually with extended hips, bent knees and pronated feet, and rounded back with arms and hands in front to avoid hyperextension.

Hypertonia exists or there may be fluctuations of tone. The rare rigid hypertonus is a 'lead pipe' rigidity. Dystonic hypertonia is present. Both show a continuous resistance to passive stretch throughout the full range of motion. Dystonic posturing can be particularly disabling, especially if combined with spasticity. Intended movement, especially in a specific task, triggers dystonia, which can be sustained or intermittent. The dystonic muscle contractions may create twisting into unusual postures and be repetitive. Rigidity is detected when movement is externally applied (Sanger et al. 2003, 2010). Arousal of emotions increases

dystonia and dyskinesia. Other triggers such as tactile stimulation, pain, constipation, and intolerance of orthoses and seating can cause dystonic posturing. Dystonic tremor may be present. Sudden flexion or extensor spasms could occur. Sudden wide opening of the mouth with spasm can take place. Sleep decreases spasms or dystonic postures. However, more severe dystonia may make sleep difficult. Deformities are less likely owing to the fluctuations of tone. There can be overlap of dystonia and dyskinesia.

Status dystonicus is rare and a medical emergency. Doctors need to educate therapists and parents well beforehand.

The athetoid dance. Some children with athetosis are unable to maintain weight on their feet and continually withdraw each foot either upwards or upwards and outwards, in an 'athetoid dance'. They may take weight on one foot while pawing or scraping the ground in a withdrawal motion with the other leg.

Paralysis of gaze movements may occur, so children with dyskinesia may find it difficult to look upwards and sometimes also to close their eyes voluntarily. Poor head control also disrupts use of the eyes. Jan et al. (2001) found severe dyskinetic eye movements in some children with dyskinesia, associated with variable and highly inefficient vision.

A dyskinetic presentation can change with time. Children may be floppy (hypotonic) in babyhood and only exhibit the involuntary movements when they reach 2 or 3 years of age. Most dystonia and dyskinesia are bilateral. There is a rare unilateral presentation. Some adults or adolescents learn to use muscle tension to control involuntary movements. Individuals may find their own ways of achieving function.

Associated impairments

1. Intelligence is frequently good and may be very high. However, intellectual impairment is still present in some individuals.
2. Hearing loss of a specific high-frequency type is associated with athetosis caused by kernicterus, although 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. Tongue thrust and drooling may be particular problems. Poor hand and arm function affects the development of self-feeding.

Ataxic cerebral palsy

The main motor characteristics are as follows.

Disturbances of balance. There is poor stabilisation of the head and trunk and of the shoulder and pelvic girdles. Unsteady gaits are present. Some children with ataxia overcompensate for this instability by having excessive balance-saving reactions in the arms. Instability is also found in children with other classifications and may be called ataxia even when the child's presentation is primarily described as dyskinetic or spastic cerebral palsy, as pure ataxia is rarely diagnosed. Pure ataxia, called 'dysequilibrium syndrome' (Neville 2000), is understood to be of genetic origin.

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. Children with ataxia often present

with all of their movements appearing disorganised or jerky. Poor fine hand movements occur when, for example, tying shoe laces, managing buttons, using a pencil, and drawing or writing.

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

Nystagmus may exist.

Associated impairments

Intellectual impairment may exist, especially in regards to non-verbal cognitive skills such as visual and perceptual difficulties.

Common features observed in all presenting types of cerebral palsies

Postural mechanisms

The classification of tone and component gross and fine motor skills has tended to obscure the fact that there are important motor features that are common to all children with a diagnosis of cerebral palsy, for example delayed motor development. However, symptoms such as spasticity, sudden spasms, and the various involuntary movements only play a part in this disturbance of development. Delayed or atypical development of the postural balance mechanisms significantly disturbs motor development. Postural mechanisms are an intrinsic part of motor skills. When they are absent or atypical, this leads to absent or atypical motor skills.

Chapters 7 and 9 discuss these aspects in detail, as they are fundamental to the framework for therapy.

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

Infantile reflexes

Besides the desirable postural mechanisms, there are reflexes that have no predilection for any specific type of cerebral palsy. These are infantile reflexes that are present in the typical newborn and that become integrated or disappear as the baby matures. In cerebral palsy infantile reflexes may still present long after the ages when they should have become integrated within the nervous system. Some older children who do not yet have postural mechanisms activate some of these reflex responses in their efforts to balance and move as the infantile reactions can be their only way to function. These reactions may be stimulated by either peripheral or cortical activations. A therapist needs to include knowledge of how their peripheral stimulation and handling might cause residual 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, excessive neck righting reflex to roll, automatic stepping when the body is fully supported in a walker, and positive supporting reaction for standing in a standing frame. Children may use extensor thrusts or Moro reactions or Startle to communicate non-verbally. Feeding reflexes disrupt independent feeding development.

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 among the infantile reflexes, whereas others argue that they are not present in the typical 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 *atypical* postural reflexes and should not be confused with the typical postural mechanisms as described by a number of neurologists and researchers (Belenkii et al. 1967; Martin 1967; Hadders-Algra and Carlberg 2008; Shumway-Cook and Woollacott 2017 among others).

The principle of treatment that therapists should follow in relation to the complicated collection of reflexes is *not* to go 'reflex hunting'. In the past, some therapists assessed directly for reflexes that might interfere with motor function and speech. However, the current approach is to examine the function of the child first and then plan therapy intervention that will support a child's functional development, which replaces the need for reflex reactions as compensation for lack of developmental motor control and function. Table 8.2 (see Chapter 8) of reflex reactions is given so that a therapist recognises any total or remnants of these reflex reactions in an individual.

Reflex reactions are not a substrate for motor control and are not reliable predictors of future motor development. Ideas on the 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. 1992; Horak 1992; Prechtl 2001; Einspieler et al. 2005). These studies lend support to 'avoid reflex hunting' expressed in this book since the first edition in 1977.

Additional impairments

Individual children, particularly those with severe cerebral palsy, may have sleep problems, fatigue, feeding problems and poor nutrition, decreased bone mineral density, musculoskeletal pain or pain from hypertonus, or severe gastro-oesophageal reflux, and are less fit than their peers without physical disabilities. Most of these problems develop in later childhood and are managed by medical consultants. Therapists are involved with feeding problems, fitness, and bone mineral density. Therapists need to be aware of any additional impairments as they may impinge on the amount of energy a child has available for both functional activities and individualised 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 energy and ability to carry out a child's home therapy. The physical demands of caregiving have been shown to negatively impact on parents' physical and psychological well-being (Ward et al. 2014). Pain and decreased bone mineral density are treated with drugs and prevented to some degree by therapists using activities involving active weight bearing (NICE 2017). See assessment of pain in 'Specialised medical treatments' in Chapter 5.

Motor delay

Cerebral palsy consists of both motor delay and motor disorder. In Chapter 8 various assessments are described that can be used to assess motor delay. There are many other conditions that may also present with similar problems in regards to the development of motor skills or of delay and disorder. All these conditions are known as *developmental disabilities* (Levitt 1984).

They may be due to the following.

Intellectual impairment. This is caused by various metabolic disorders, chromosome anomalies, leukodystrophies, microcephaly and other abnormalities of the skull and brain, endocrine disorders, and the causes of brain damage given for the cerebral palsies. The latest version of the Diagnostic and Statistical Manual of Mental Health Disorders (DSM-V) includes functional difficulties as a criterion for a diagnosis of intellectual disability alongside cognitive ability as measured by an individual's IQ level.

Deprivation of stimulation associated with social, economic, or severe emotional problems.

Malnutrition alone, but usually together with deprived environments.

Once malnutrition is treated, lack of normal stimulation may still delay the child's development.

The presence of non-motor impairments. This may influence motor development and lead to delay, for example severe visual impairments. Children with delay in any developmental area are more likely to have an associated delay in motor development.

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

Principles of treatment and organisation of treatment. These will be similar to those discussed in Chapters 1, 2, 3, and 4. *Specific problems* in the conditions above are considered in other publications (Levitt 1984; Campbell et al. 2011; Rodger and Kennedy-Behr 2017).

Summary

This chapter provides basic information for planning treatment and management.

1. The child with cerebral palsy has abilities and disabilities primarily in motor control and function, but may have individual associated difficulties in the areas of communication, cognition, sensory, and perceptual functions.
2. 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.
3. There is an interaction between the communication, intellectual, sensory, perceptual, and motor functions. Therapists therefore consider the influence of associated abilities and disabilities on therapy programmes.
4. Treatment is aimed at development and support of gross motor and fine motor functions to enable engagement in activities and increase participation.
5. Specific attention will need to be paid to the following:
   a. postural mechanisms of stability and balance
   b. voluntary movement patterns (synergies) of body, limbs, and hands
   c. strengthening for weakness of various kinds
   d. minimising hypertonicity, hypotonicity, and involuntary movements
   e. improving postural alignments and patterns of gait
   f. improving ranges of motion of muscles, joints, and soft tissues.

   Therapists also need to reflect on the relationships of these aspects.
6. Therapy programmes should not have a strict adherence to specific diagnostic classifications as there can be clinical changes. Aetiology may not always influence therapy.
7. Therapy includes:
   a. Assessment and management based on the perspectives of an individual, the family, and others involved with that individual. Consideration of attitudes in the family and society that facilitate or act as barriers to an individual's function and participation.
   b. Focus on developmental therapy, minimising impairments *within the functions*.
   c. Therapists always recognising the emerging functional *abilities and whole functions* within each child's development.
   d. Assessment and prevention of secondary and increasing impairments.

   e. Encouragement of the positive attributes of individuals with cerebral palsy and of their families.
   f. Promoting positive motor experiences as key for motivating the best therapy.
8. Typical developmental schedules are only guides and need to be carefully adapted.
9. 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.
10. Therapists need to integrate learning principles that encompass emotional, cultural, and social issues.
11. Treatment and management need to commence as early as possible for parental support, parent–child relationships, and to promote a child's activities as well as minimise any musculoskeletal problems.

The model suggested by the ICF provides a framework for understanding disability and health in the twenty-first century. It acts as a general guide to assessment and planning of therapy and management, which matches many discussions in this book.