المشاكل التطورية والأطفال ذوي الاحتياجات الخاصة

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Developmental problems and the child with special needs

Features of developmental problems and the child with special needs are:

- developmental problems present in the perinatal period and throughout childhood
- with developmental delay, the difference with their peers increases as the child gets older
- cerebral palsy is the most common cause of motor impairment in children
- in autism spectrum disorder, there is abnormal development of social and communication skills
- attention deficit hyperactivity disorder (ADHD) needs to be differentiated from normal, boisterous children
- early detection of severe impairment of hearing or vision is important to minimize its detrimental effect on development
- their medical, social, emotional and educational requirements are complex
- are looked after by local multidisciplinary child development services.

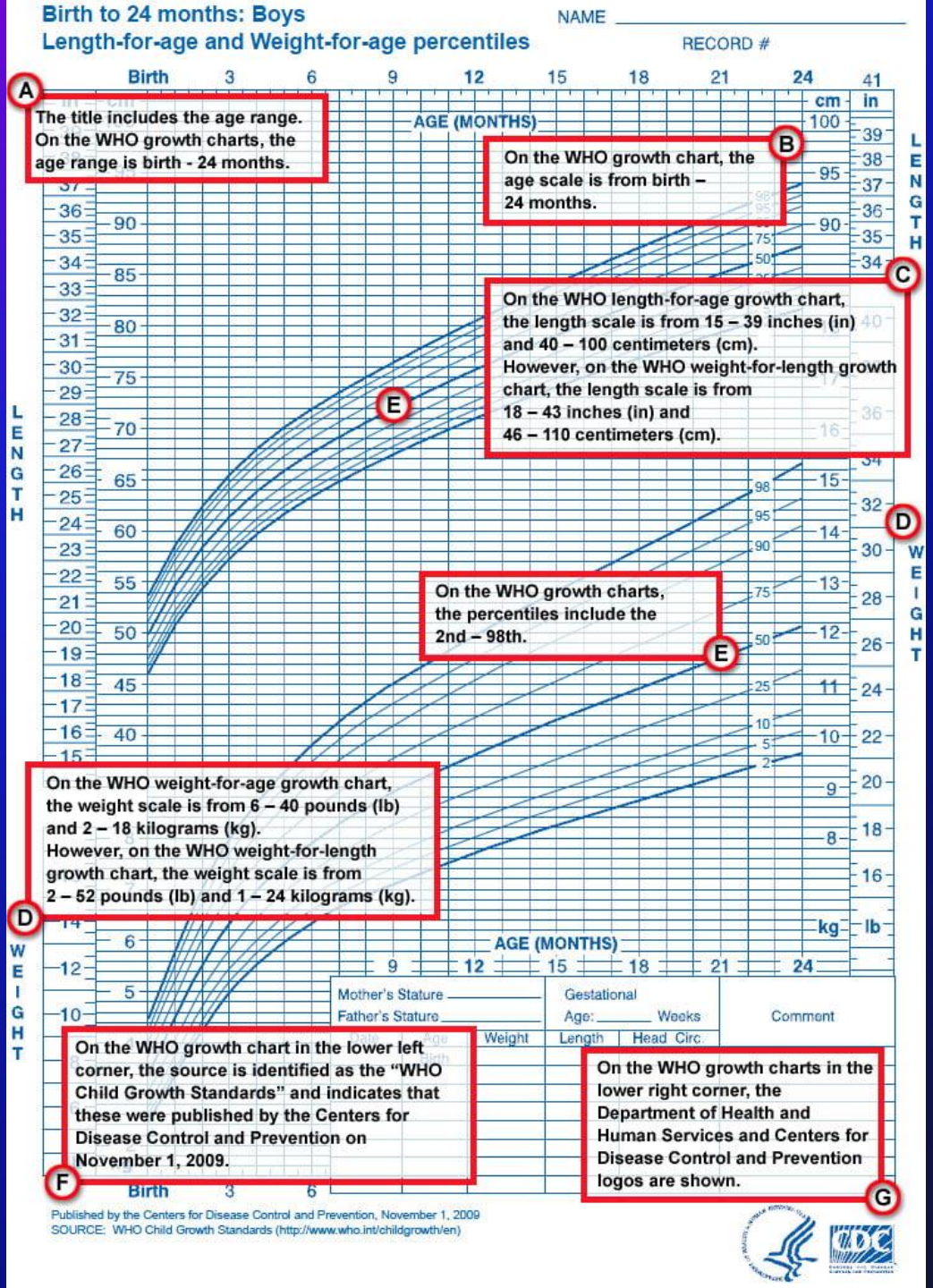
Any child whose development is delayed or disordered needs assessment to determine the cause and management. Neurodevelopmental problems present at all ages, with an increasing number now recognized antenatally (Table 4.1). Many are identified in the neonatal period because of abnormal neurology or dysmorphic features. During infancy and early childhood, problems often present at an age when a specific area of development is most rapid and prominent (i.e. motor problems during the first 18 months of age, speech and language problems between 18 months and 3 years, and social and communication disorders between 2–4 years of age). Abnormal development may be caused not only by neurodevelopmental disorders (Table 4.2) but also by ill health or if the child's physical or psychological needs are not met. When carrying out a clinical examination on a young child with a possible developmental problem:

- ask the parent what the child's abilities are. Start at a level below what a child of that age is likely to be able to do to retain confidence of the parent and child
- *observe* the child from the first moment seen
- make it fun. The assessment should be perceived as a game by the child
- toys to use are cubes, a ball, car, doll, pencil, paper, pegboard, miniature toys, picture book. Adapt their use to the child
- formulate a developmental picture in terms of gross motor; vision and fine motor; hearing, speech and language; and social, emotional and

behaviour. As you become more confident, you will screen all these skills simultaneously

- assess the child to a short level above what they appear able to do in order to establish their ceiling of skill for each developmental area
- remember to adjust developmental expectations for prematurity
- at the end of developmental screening you should be able to describe what a child is able to do and what the child cannot do, if the abilities are within normal limits for age and, if not, which developmental fields are outside the normal range

- clinical signs to look for that may aid diagnosis or guide investigation are:
  - patterns of growth: height, weight, head circumference with centile plotting
  - dysmorphic features: face, limbs, body proportions, cardiac, genitalia
  - skin: neurocutaneous stigmata, injuries, cleanliness


Birth to 24 months: Boys
Length-for-age and Weight-for-age percentiles
NAME
RECORD #
AGE (MONTHS)
LENGTH
WEIGHT
cm
in
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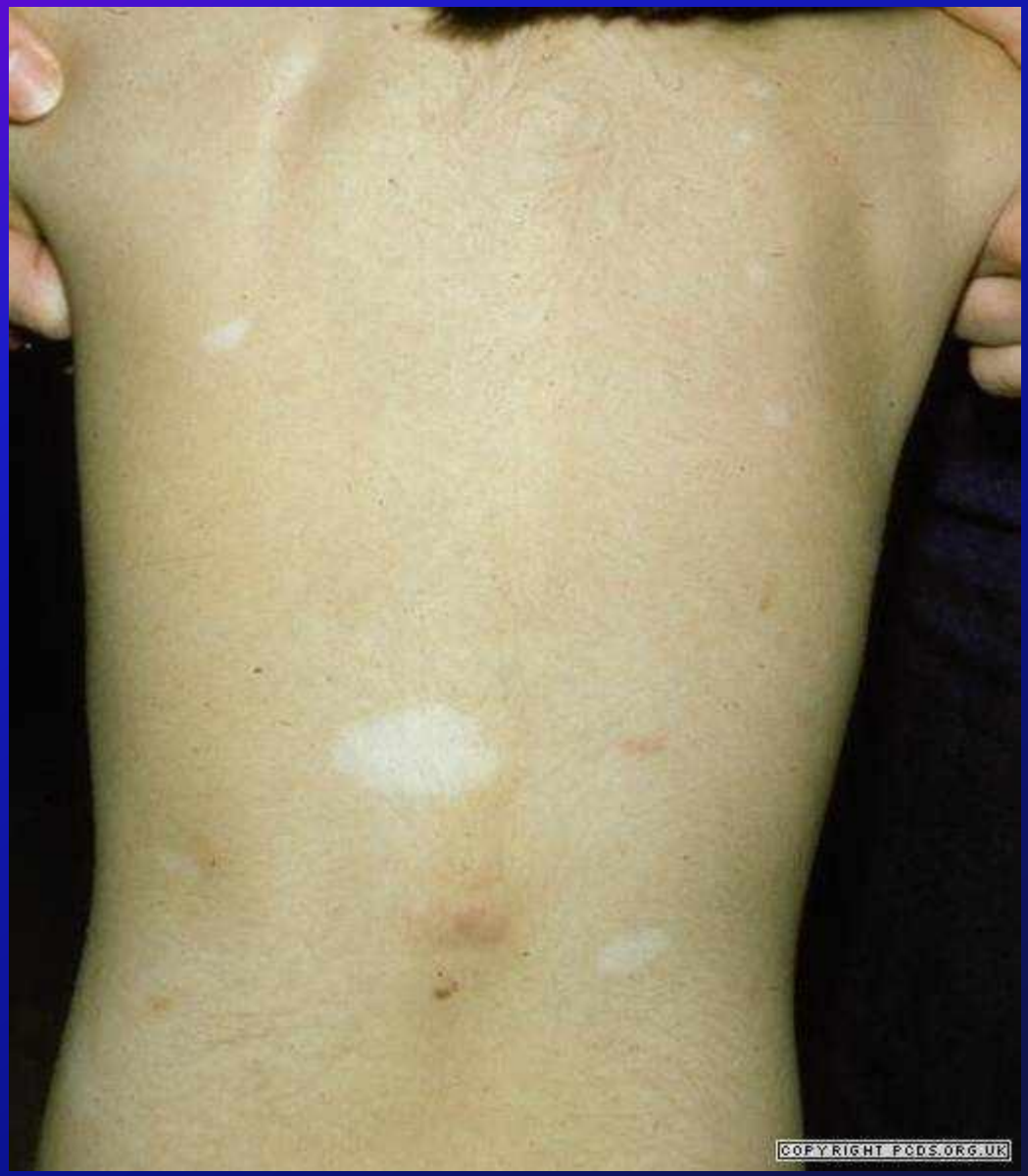
A
The title includes the age range. On the WHO growth charts, the age range is birth - 24 months.
B
On the WHO growth chart, the age scale is from birth – 24 months.
C
On the WHO length-for-age growth chart, the length scale is from 15 – 39 inches (in) and 40 – 100 centimeters (cm). However, on the WHO weight-for-length growth chart, the length scale is from 18 – 43 inches (in) and 46 – 110 centimeters (cm).
D
On the WHO weight-for-age growth chart, the weight scale is from 6 – 40 pounds (lb) and 2 – 18 kilograms (kg). However, on the WHO weight-for-length growth chart, the weight scale is from 2 – 52 pounds (lb) and 1 – 24 kilograms (kg).
E
On the WHO growth charts, the percentiles include the 2nd – 98th.
F
On the WHO growth chart in the lower left corner, the source is identified as the "WHO Child Growth Standards" and indicates that these were published by the Centers for Disease Control and Prevention on November 1, 2009.
G
On the WHO growth charts in the lower right corner, the Department of Health and Human Services and Centers for Disease Control and Prevention logos are shown.
Mother's Stature
Father's Stature
Gestational Age: Weeks
Comment
Weight
Length
Head Circ.
Published by the Centers for Disease Control and Prevention, November 1, 2009
SOURCE: WHO Child Growth Standards (http://www.who.int/childgrowth/en)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- central nervous system examination: abnormal posture/symmetry, wasting, tone and power, deep tendon reflexes, clonus, plantar responses, cranial nerves
- cardiovascular examination: abnormalities are associated with many dysmorphic syndromes
- visual function and ocular abnormalities
- hearing: by questioning parents about hearing and language development and checking if neonatal hearing screening was done
- patterns of mobility, dexterity, hand dominance, communication and social skills, general behaviour
- cognition.

Table 4.1 Features that may suggest neurodevelopmental concerns by age

Prenatal	Positive family history, e.g. affected siblings or family members; ethnicity, e.g. Tay–Sachs disease in Jewish parents
	Antenatal screening tests, e.g. ultrasound including nuchal thickness, triple blood test or non-invasive prenatal testing (NIPT, cell-free DNA testing of fetal cells from maternal blood) for conditions such as Down syndrome; neural tube defects, e.g. spina bifida and hydrocephalus. Amniocentesis for suspected genetic disorders
Perinatal	Following birth asphyxia/neonatal encephalopathy
	Preterm infants with intraventricular haemorrhage/periventricular leucomalacia, post-haemorrhagic hydrocephalus
	Dysmorphic and neurocutaneous features
	Abnormal neurological behaviour – tone, feeding, movement, seizures, visual inattention
Infancy	Global developmental delay
	Delayed or asymmetric motor development
	Neurocutaneous and dysmorphic features (cataracts)
	Vision or hearing concerns by parents or after screening

Preschool	Speech and language delay
	Abnormal gait, clumsy motor skills
	Poor social communication skills
	Behaviour – stereotypical, overactivity, inattention
School age	Problems with balance and coordination
	Learning difficulties
	Attention control
	Hyperactivity
	Specific learning difficulties, e.g. dyslexia, dyspraxia
	Social communication difficulties
Any age	Acquired brain injury, e.g. after meningitis, head injury
	Loss of skills

Many examination findings can be predicted from *observation* of functional skills and behaviour.

Many parental concerns about their child's development are found to be variations of normal, in which case the parents should be reassured. If in doubt, observe the child's progress over a period of time.

Table 4.2 Conditions that cause abnormal development and learning difficulty

Prenatal	
Genetic	Chromosome/DNA disorders, e.g. Down syndrome, fragile X syndrome, chromosome microdeletions or duplications Cerebral dysgenesis, e.g. microcephaly, absent corpus callosum, hydrocephalu neuronal migration disorder
Cerebrovascular	Stroke – haemorrhagic or ischaemic
Metabolic	Hypothyroidism, phenylketonuria
Teratogenic	Alcohol and drug abuse
Congenital infection	Rubella, cytomegalovirus, toxoplasmosis, HIV
Neurocutaneous syndromes	Tuberous sclerosis, neurofibromatosis, Sturge–Weber, Ito syndrome

Perinatal	
Extreme prematurity	Intraventricular haemorrhage/periventricular leucomalacia
Birth asphyxia	Hypoxic-ischaemic encephalopathy
Metabolic	Symptomatic hypoglycaemia, hyperbilirubinemia
Postnatal	
Infection	Meningitis, encephalitis
Anoxia	Suffocation, near drowning, seizures
Trauma	Head injury – accidental or non-accidental
Metabolic	Hypoglycaemia, inborn errors of metabolism.
Cerebrovascular	Stroke
Nutritional deficiency	Maternal deficiency (breast fed), food intolerances, restrictions

Other Unknown (about 25%): chronic illness, physical abuse, emotional neglect	

Note: The site and severity of brain damage influence the clinical outcome, i.e. whether specific or global developmental delay, learning and/or physical disability.

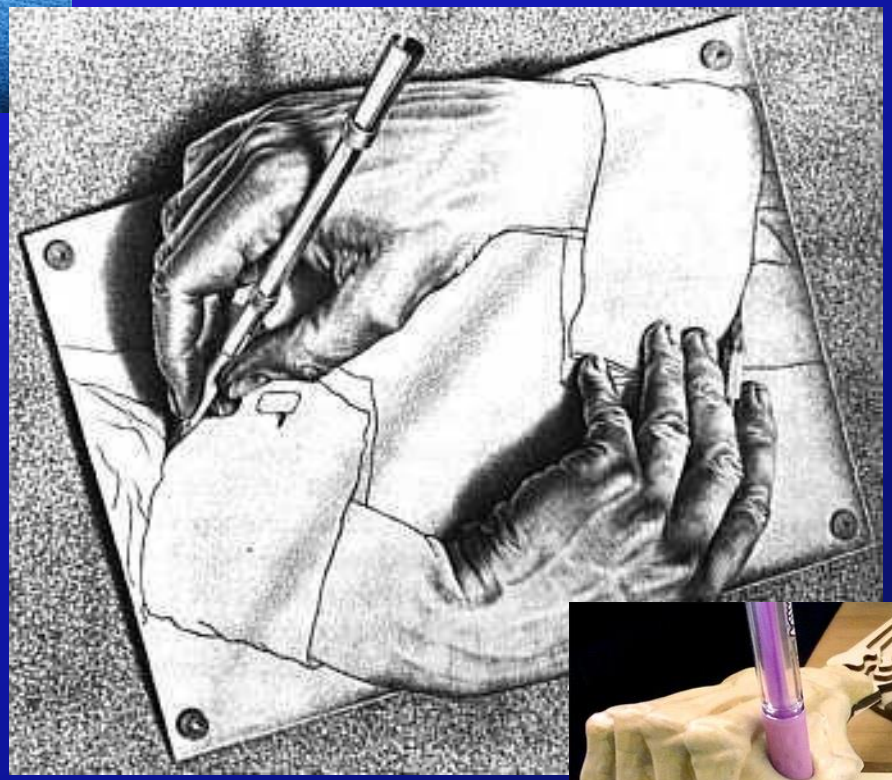
Abnormal development – key concepts

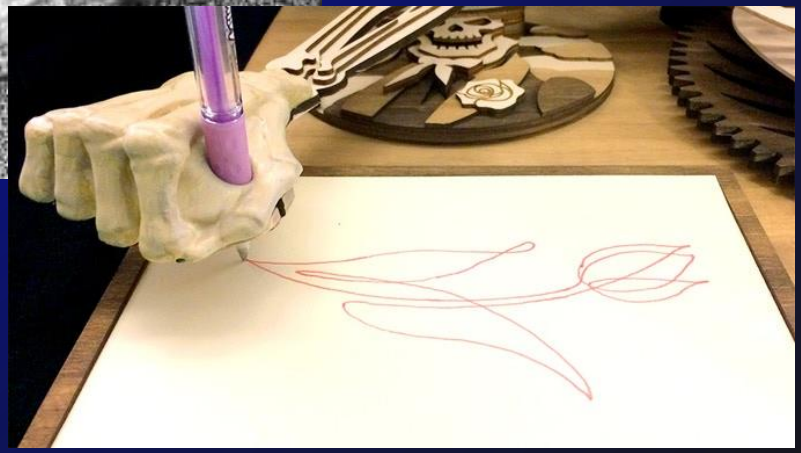
The terminology can be confusing, but:

- *delay* – implies slow acquisition of all skills (global delay) or of one particular field or area of skill (specific delay), particularly in relation to developmental problems in the 0–5-year age group
- *learning difficulty* – used in relation to children of school age and may be cognitive, physical, both, or relate to specific functional skills
- *disorder* – maldevelopment of a skill.

The following are agreed definitions:

- *impairment* – loss or abnormality of physiological function or anatomical structure
- *disability* – any restriction or lack of ability due to the impairment
- *disadvantage* – this results from the disability, and limits or prevents fulfilment of a normal role. It is situationally specific; a child with a learning disability may, for example, be a good skier or enjoy swimming.



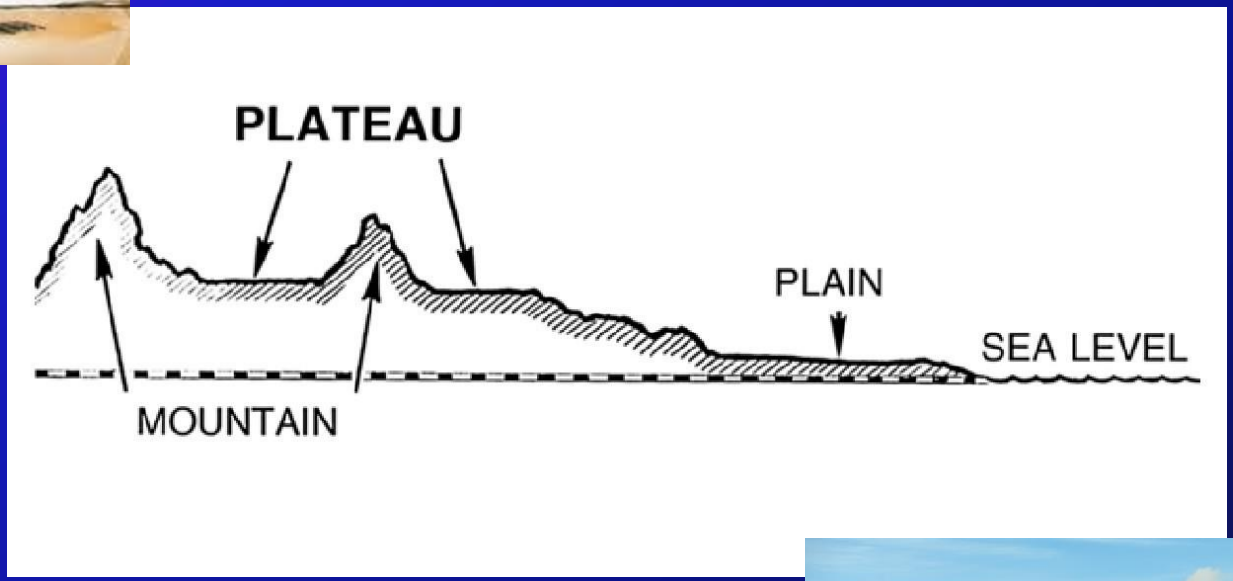


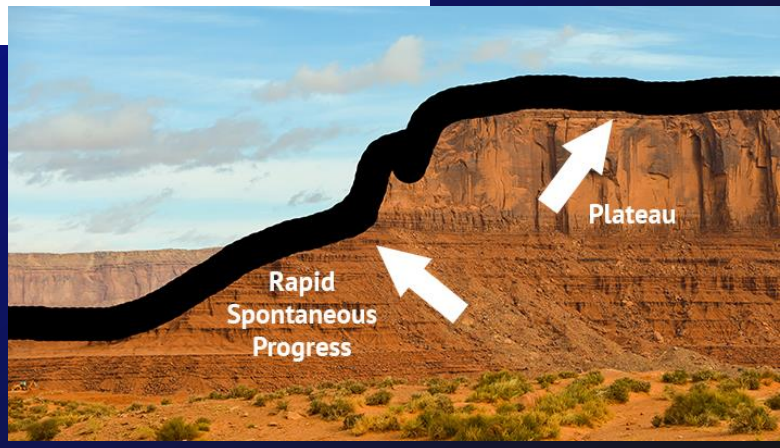
The term 'handicap' is now discouraged as it can imply a person deserves pity. Difficulty and disability are often used interchangeably, but difficulty is used particularly in an educational context. Impairment is now generally used instead of disability when describing problems such as visual impairment or hearing impairment.

The *pattern* of abnormal development (global or specific) can be categorized as (Fig. 4.1):

- slow but steady
- plateau effect
- showing regression
  - acute regression following acute brain injury with subsequent slow recovery but not to normal levels (partial recovery) or slow regression as with neurodegenerative disorders.


SLOW AND STEADY


PLATEAU
PLAIN
SEA LEVEL
MOUNTAIN


Plateau
Rapid
Spontaneous
Progress

The *severity* can be categorized as:

- mild
- moderate
- severe
- profound.

Other features of developmental delay are:

- the gap between normal and abnormal development becomes greater with increasing age, and therefore becomes more apparent over time (Fig. 4.2)

- it may be the presentation of a wide variety of underlying conditions (Table 4.2)
- the site and severity of brain damage influences the clinical outcome, i.e. whether there will be specific or global developmental delay, learning and/or physical disability
- it may be genetic, with important implications for the family
- there is a wide age band across which it can be normal to achieve a developmental skill
- limit ages denote beyond the normal range.

The choice of investigations to identify the cause is influenced by the child's age, the history and clinical findings (Table 4.3). In some children, no cause can be identified even after extensive investigation.

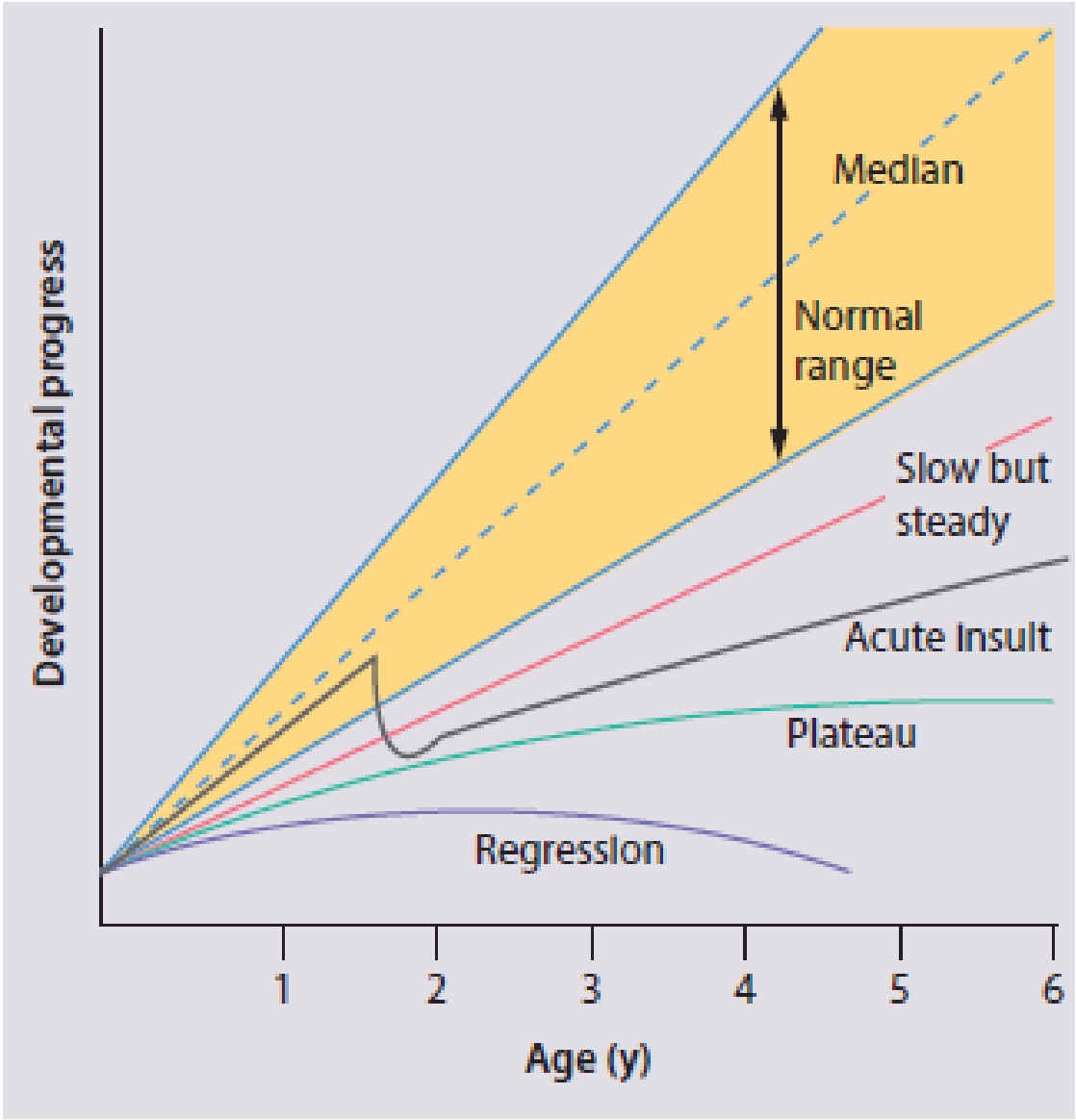


Figure 4.1 Patterns of abnormal development. These may be slow but steady, plateau, regression. They may follow an acute injury.

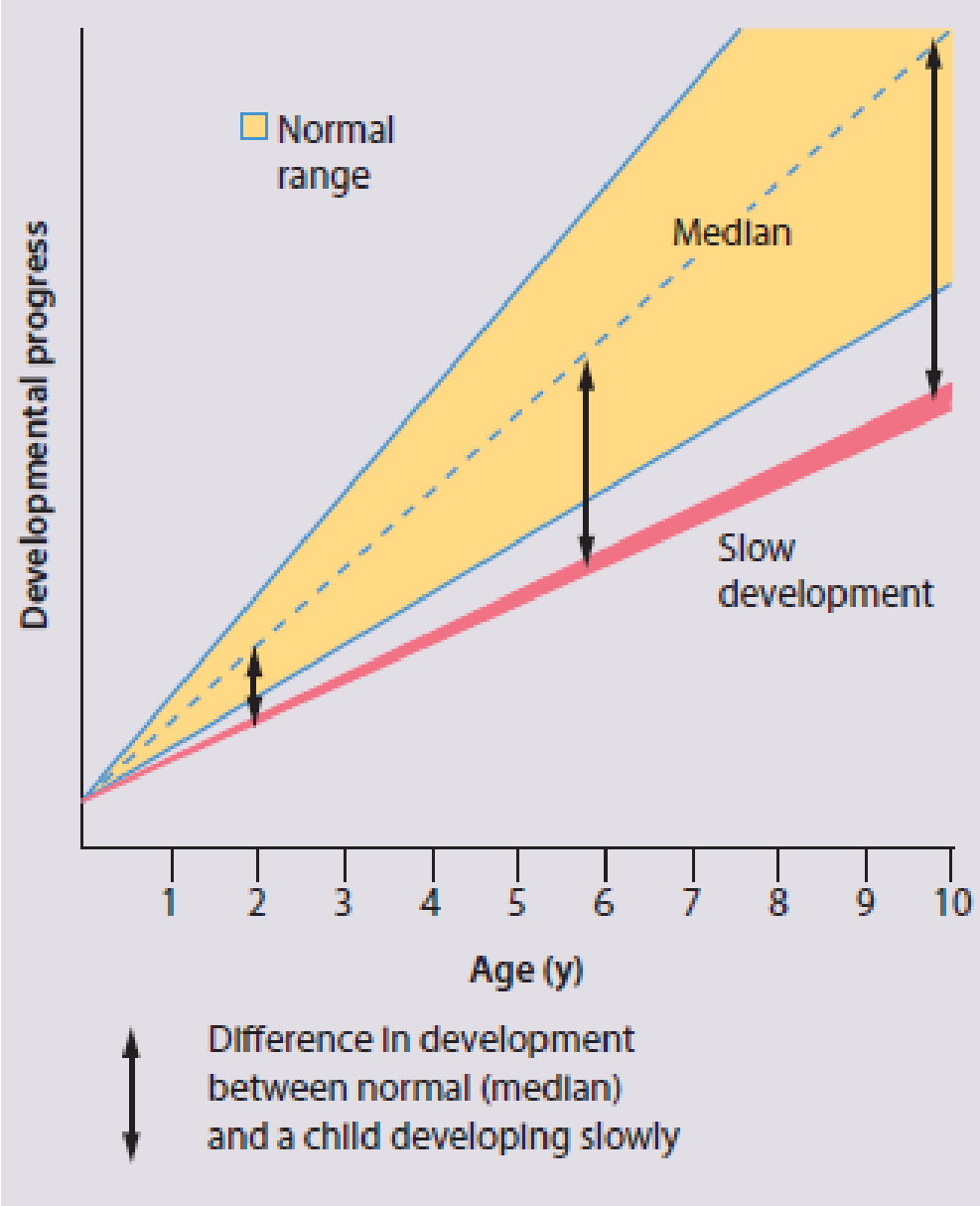


Figure 4.2 For children with abnormal development, the gap between their abilities and what is normal widens with age.

Summary

Disordered development

- Incorporates global and specific delay or disorder, learning difficulty, impairment and disability.
- Varies in pattern of progression and severity.
- Becomes more apparent with age.

Language Delay

Speech Delay

Motor Skill Issues

Intellectual Development

Emotional Development

Developmental Delay

Social Skills Development

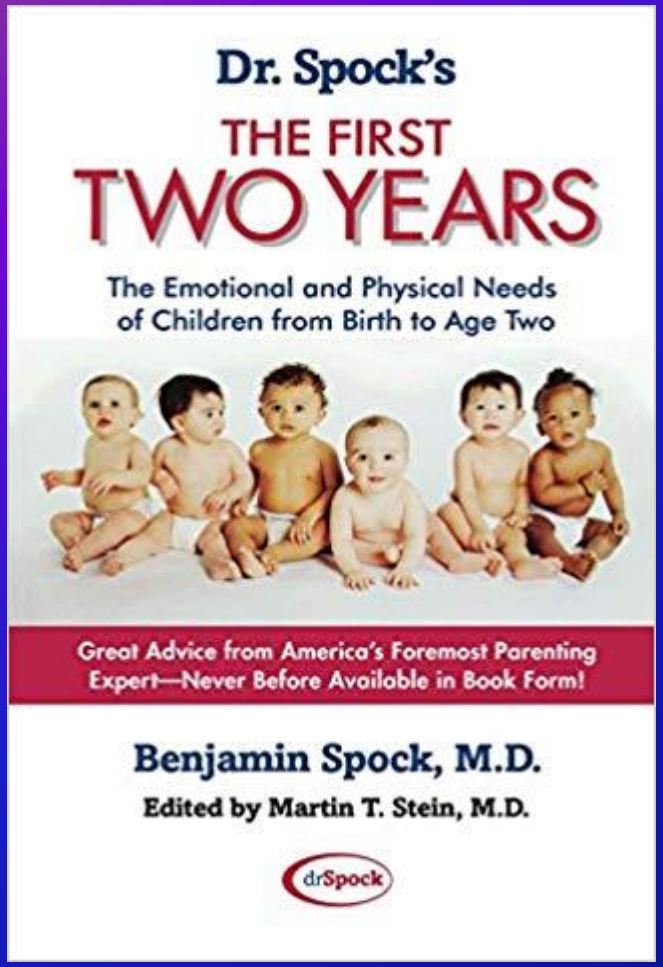
Movement Problem

Cognitive Development

Developmental delay

Global developmental delay (also called early developmental impairment) implies delay in acquisition of all skill fields (gross motor, vision and fine motor, hearing and speech, language and cognition, social/emotional and behaviour). It usually becomes apparent in the first 2 years of life. Global developmental delay is likely to be associated with cognitive difficulties, although these may only become apparent several years later. The presence of global developmental delay should always generate investigation into a possible cause such as those listed in Table 4.2. When children become older and the clinical picture is clearer, it is more appropriate to describe the individual difficulties such as learning disability, motor disorder and communication difficulty, rather than using the term global developmental delay.

Specific developmental impairment is when one field of development or skill area is more delayed than others. It may also be developing in a disordered way.



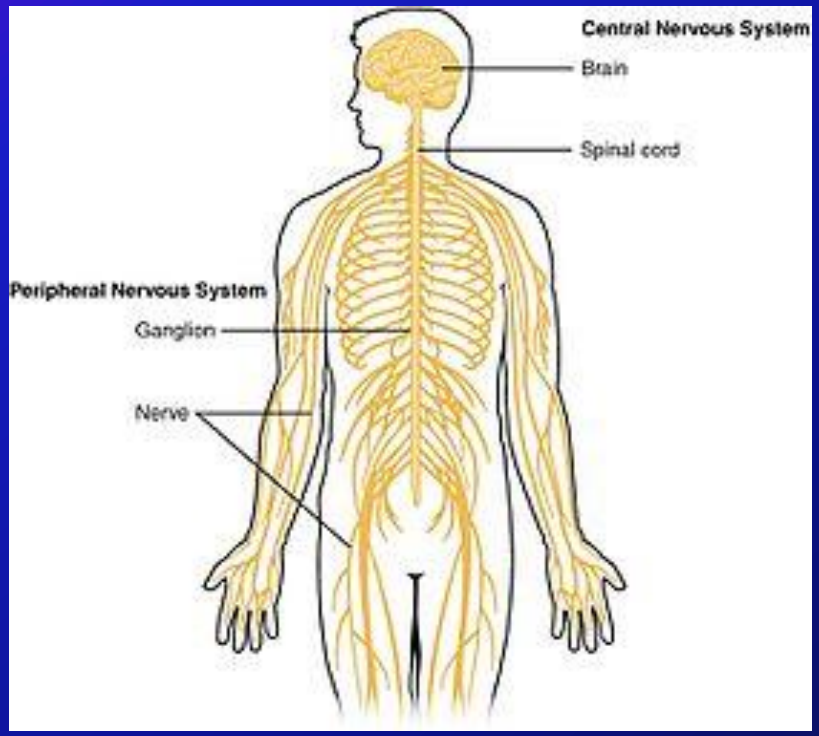
Global developmental delay usually presents in the first 2 years of life.

Abnormal motor development

This may present as a delay in acquisition of motor skills, e.g. head control, rolling, sitting, standing, walking or as problems with balance, an abnormal gait, asymmetry of hand use, involuntary movements or rarely loss of motor skills. Concern about motor development usually presents between 3 months–2 years of age when acquisition of motor skills is occurring most rapidly. Examination may reveal underlying abnormal motor signs.

Causes of abnormal motor development include:

- central motor deficit, e.g. cerebral palsy (CP)
- congenital myopathy/primary muscle disease
- spinal cord lesions, e.g. spina bifida
- global developmental delay, as in many syndromes, or of unidentified cause.



As hand dominance is not acquired until 1–2 years of age or later, asymmetry of motor skills during the first year of life is always abnormal and may suggest an underlying hemiplegia.

Late walking (>18 months old) may be caused by any of the aforementioned causes but also needs to be differentiated from children who display the normal locomotor variants of bottom-shuffling or commando crawling (see Ch. 3) where walking occurs later than with crawlers, and from children with joint hypermobility who may also achieve walking later than average.

Concern about abnormal motor development needs assessment by a neurodevelopmental paediatrician and physiotherapist. Ongoing physiotherapy input and subsequent involvement of an occupational therapist are also likely to be needed.

Table 4.3 Investigations or assessment to consider for developmental delay

Cytogenetic	Comparative genomic hybridization microarray or chromosome karyotype[a]
	Fragile X analysis[a]
	DNA fluorescence in situ hybridization analysis, e.g. for chromosome 7, 15, and 22 deletions; telomere screening; whole-exome sequencing
Metabolic	Thyroid function tests, liver function tests, bone chemistry, urea and electrolytes, plasma amino acids[a], blood film
	Creatine kinase, blood lactate, very long-chain fatty acids, ammonia, blood gases, white cell (lysosomal) enzymes, urine amino and organic acids, urine mucopolysaccharides (GAG), and oligosaccharide screen, urine reducing substances, lead levels, urate, ferritin, biotinidase, vitamin B6 and B12
	Maternal amino acids for raised phenylalanine

Infection	Congenital infection screen for cytomegalovirus, etc.
Imaging	Cranial ultrasound in newborn
	CT and MRI brain scans
	Skeletal survey, bone age
Neurophysiology	EEG – for seizures and can be diagnostic for specific neurological disorders and syndromes
	Nerve conduction studies, electromyogram, visual evoked potentials, electroretinogram
Histopathology/ histochemistry	Nerve, skin and muscle biopsy

Other	Hearing[a]
	Vision[a]
	Clinical genetics
	Cognitive and behavioural assessment (clinical and educational psychologist)
	Therapy assessment – physiotherapy, occupational therapy, speech and language therapy
	Child psychiatry
	Dietician
	Nursery/school reports[a]

Cerebral Palsy

"Cerebral"

refers to the cerebrum, which is the affected area of the brain

"Palsy"

refers to the brain injury's resultant disorder of movement

Cerebral palsy (CP) is a group of motor conditions that causes physical disability in development, primarily in various areas of body movement.

It is caused by damage to the developing brain's motor control centers

Cerebral palsy

CP is difficult to define, but the international group for Surveillance of Cerebral Palsy in Europe defines it as an umbrella term for a permanent disorder of movement and/or posture and of motor function due to a non-progressive abnormality in the developing brain.

The motor disorders of CP are often accompanied by disturbances of cognition, communication, vision, perception, sensation, behaviour, seizure disorder and secondary musculoskeletal problems. Although the causative lesion is non-progressive and damage to the brain is static, clinical manifestations emerge over time, reflecting the balance between normal and abnormal cerebral maturation. Motor dysfunction is usually evident early, often from birth. If the brain injury occurs after the age of 2 years, it is diagnosed as acquired brain injury.

CP is the most common cause of motor impairment in children, affecting about 2 per 1000 live births.

Causes

About 80% of CP is antenatal in origin due to cerebrovascular haemorrhage or ischaemia, cortical migration disorders or structural maldevelopment of the brain during gestation. Some of these problems are linked to gene deletions. Other antenatal causes are genetic syndromes and congenital infection.

Only about 10% of cases are thought to be due to hypoxic-ischaemic injury before or during delivery and this proportion has remained relatively constant over the last decade. About 10% are postnatal in origin.

Preterm infants are especially vulnerable to brain damage from periventricular leukomalacia secondary to ischaemia and/or severe intraventricular haemorrhage and venous infarction. The improved survival of extremely preterm infants has been accompanied by an increase in survivors with CP, although the number of such children is relatively small.

Postnatal causes are meningitis/encephalitis/encephalopathy, head trauma from accidental or non-accidental injury, symptomatic hypoglycaemia, hydrocephalus and hyperbilirubinemia.

MRI brain scans may assist in identifying the cause of the CP, in directing further investigations and in supporting explanations to the parents, but is not required to make the diagnosis.

Clinical presentation

Many children who develop CP will have been identified as being at risk in the neonatal period. Early features of CP are:

- abnormal limb and/or trunk posture and tone in infancy with delayed motor milestones (Fig. 4.3); this may be accompanied by slowing of head growth
- feeding difficulties, with oromotor incoordination, slow feeding, gagging and vomiting
- abnormal gait once walking is achieved
- asymmetric hand function before 12 months of age.

In CP, primitive reflexes, which facilitate the emergence of normal patterns of movement and which need to disappear for motor development to progress, may persist and become obligatory (see Ch. 3).

The diagnosis is made by clinical examination, with particular attention to assessment of posture and the pattern of tone in the limbs and trunk, hand function and gait.

CP is now categorized according to neurological features as:

- spastic: bilateral, unilateral, not otherwise specified (90%)
- dyskinetic (6%)
- ataxic (4%)
- other.

The gross motor function level (functional ability) is described using the Gross Motor Function Classification System (Table 4.4).

Table 4.4 Gross motor function classification system (GMFCS)

Level I	Walks without limitations
Level II	Walks with limitations
Level III	Walks using a handheld mobility device
Level IV	Self-mobility with limitations; may use powered mobility
Level V	Transported in a manual wheelchair

Note: See http://www.canchild.ca/en/measures/gmfcs.asp for further details.

In the past, the description was based on the parts of the body affected (hemiplegia, quadriplegia, diplegia).

For children with high-risk factors for brain damage such as significant prematurity or those with difficulties around the time of birth, a formal standardized assessment of general movements may identify at a very young age those at greater risk of developing CP. It is a specialized assessment usually performed by a trained therapist or clinician.

Spastic cerebral palsy

In this type, there is damage to the upper motor neurone (pyramidal or corticospinal tract) pathway. Limb tone is persistently increased (spasticity) with associated brisk deep tendon reflexes and extensor plantar responses. The tone in spasticity is velocity dependent, so the faster the muscle is stretched the greater the resistance it will have. This elicits a dynamic catch, which is the hallmark of spasticity. The increased limb tone may suddenly yield under pressure in a 'clasp knife' fashion. Limb involvement is increasingly described as unilateral or bilateral to acknowledge asymmetrical signs. Spasticity tends to present early and may even be seen in the neonatal period. Sometimes there is initial hypotonia, particularly of the head and trunk. There are three main types of spastic CP:

- unilateral (*hemiplegia*) – unilateral involvement of the arm and leg. The arm is usually affected more than the leg, with the face spared. Affected children often present at 4–12 months of age with fisting of the affected hand, a flexed arm, a pronated forearm, asymmetric reaching, hand function or toe pointing when lifting the child. Subsequently, a tiptoe walk (toe–heel gait) on the affected side may become evident. Affected limbs may initially be flaccid and hypotonic, but

increased tone soon emerges as the predominant sign. The medical history may be normal, with an unremarkable birth history and no evidence of hypoxic-ischaemic encephalopathy giving rise to the possibility of a prenatal cause, which is often silent. In some children, the condition is caused by neonatal stroke. More severe vascular insults may cause a hemianopia (loss of half of visual field) of the same side as the affected limbs

- bilateral (*quadriplegia*) – all four limbs are affected, often severely. The trunk is involved with a tendency to opisthotonus (extensor posturing), poor head control and low central tone (Fig. 4.4). This more severe form of CP is often associated with seizures, microcephaly and moderate or severe intellectual impairment. There may have been a history of perinatal hypoxic-ischaemic encephalopathy

- bilateral (*diplegia*) – all four limbs, but the legs are affected to a much greater degree than the arms, so that hand function may appear to be relatively normal. Motor difficulties in the arms are most apparent with functional use of the hands. Walking is abnormal. Diplegia is one of the patterns associated with preterm birth due to periventricular brain damage. The MRI brain scan may show periventricular leukomalacia.

Normal and abnormal motor development

Normal motor development

– pushes up on arms
– holds head up

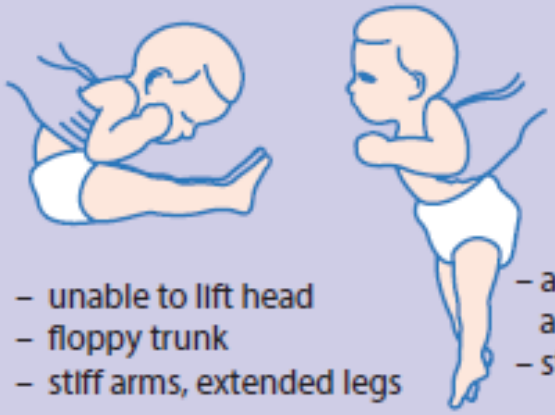
– sits with support
– holds head up
– rounded back

Median age	Limit age
1½ months	3 months
3 months	6 months

Abnormal motor development

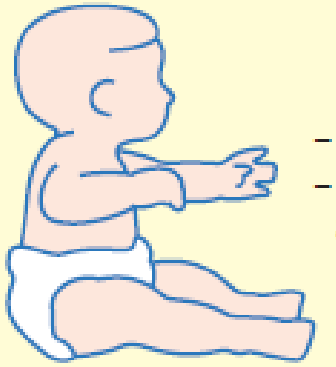
– unable to lift head or push up on arms
– stiff extended legs

– pushing back with head
– constantly fisted hand and stiff leg on one side
– difficulty moving out of this position



– unable to lift head
– floppy trunk
– stiff arms, extended legs

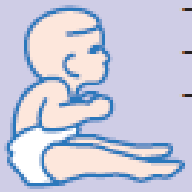
– arms flexed and held back
– stiff, crossed legs



– sits without support
– arms free to reach and grasp

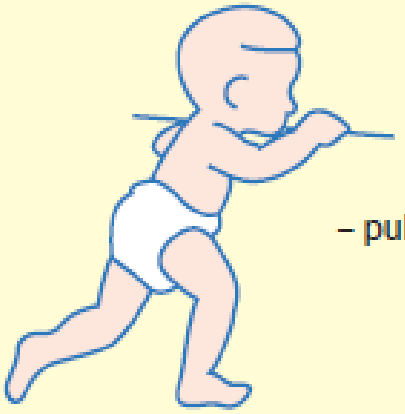
6 months 9

– rounded back
– poor use of arms for play
– stiff legs, pointed toes



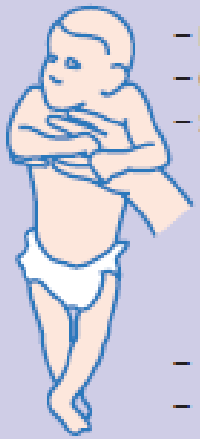
– poor head control
– difficulty getting arms forward
– arches back – stiff legs

– poor ability to lift head and back
– will not take weight on legs

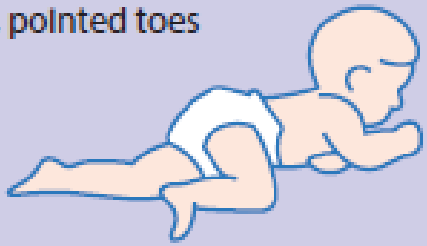


– pulls to stand

9 months 13



– not interested in weight bearing
– difficulty in pulling to stand
– stiff legs, pointed toes



– cannot crawl on hands and knees
– may use only one side of body to move

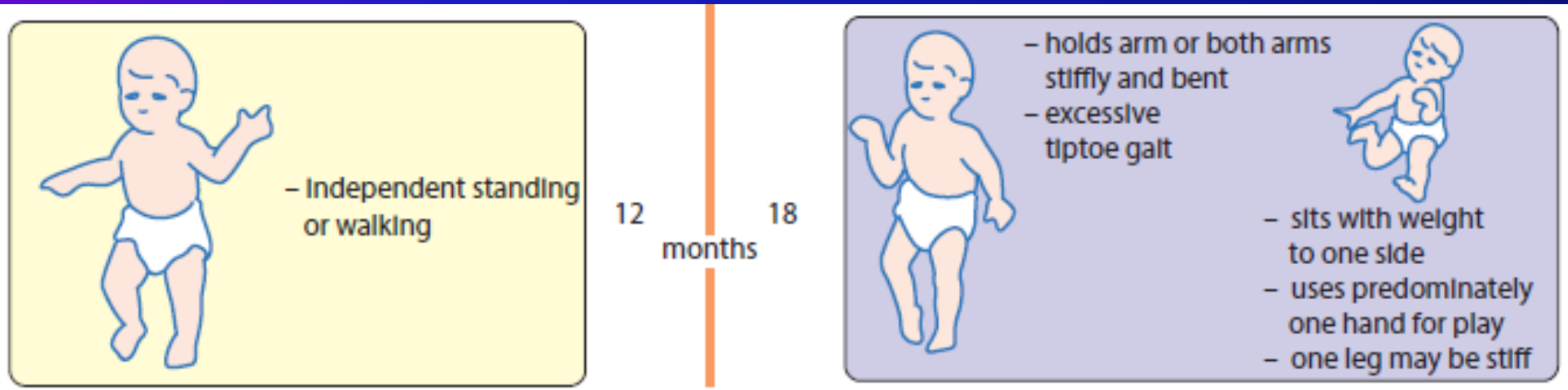


Figure 4.3 Normal motor milestones and patterns of abnormal motor development. Cerebral palsy (hemiplegia or quadriplegia) is the most common cause of developmental problems. (Adapted from Pathways Awareness Foundation, Chicago, IL.; see also https://pathways.org/)

Dyskinetic cerebral palsy

Dyskinesia refers to movements that are involuntary, uncontrolled, occasionally stereotyped and often more evident with active movement or stress. Muscle tone is variable and primitive motor reflex patterns predominate. May be described as:

- chorea – irregular, sudden and brief non-repetitive movements
- athetosis – slow writhing movements occurring more distally such as fanning of the fingers
- dystonia – simultaneous contraction of agonist and antagonist muscles of the trunk and proximal muscles often giving a twisting appearance.

Intellect may be relatively unimpaired. Affected children often present with floppiness, poor trunk control and delayed motor development in infancy. Abnormal movements may only appear towards the end of the first year of life. The signs are due to damage or dysfunction in the basal ganglia or their associated pathways (extra-pyramidal). In the past, the most common cause was hyperbilirubinemia (kernicterus) due to rhesus disease of the newborn but it is now hypoxic-ischaemic encephalopathy at term. The MRI brain scan will often show bilateral changes predominantly in the basal ganglia.

Ataxic (hypotonic) cerebral palsy

Most are genetically determined. When due to acquired brain injury (cerebellum or its connections), the signs occur on the same side as the lesion but are usually relatively symmetrical. There is early trunk and limb hypotonia, poor balance and delayed motor development. Incoordinate movements, intention tremor and an ataxic gait may be evident later.

The different types of CP are summarized in Fig. 4.5.

الأمراض العصبية العضلية

	عصبون محرك علوي	نوى قاعدية	مخيخ	خلايا القرن الأمامي	عصب محيطي	عضلية
قوة	↓	↔	↔	↓	↓	↓
مقوية	عادة تشنج	صمل أو ↓	↓	↓	↓	↓ ↔
توافق	↓	↓	↓	↔	↔	↔
حركات لا إرادية	—	رقص كنع عسرة مقوية	رجفان قصدي	تحزمات	—	—
منعكسات وترية	↑	↔	↓	غائبة أو ↓	غائبة أو ↓	↓
بابنسكي	+	-	-	-	-	-
نقص الحس	عادة +	-	-	-	+	-

Management

Parents should be given details of the diagnosis as early as possible, but prognosis is difficult during infancy until the severity and pattern of evolving signs and the child's developmental progress have become clearer over several months or years of life. Children with CP are likely to have a wide range of associated medical, psychological and social problems, making it essential to adopt a multidisciplinary approach to assessment and management, as described later in this chapter.

There are recently developed novel treatments for treating hypertonia in CP such as botulinum toxin injections to muscles, selective dorsal rhizotomy (a proportion of the nerve roots in the spinal cord are selectively cut to reduce spasticity), intrathecal baclofen (a skeletal muscle relaxant) and deep brain stimulation of the basal ganglia.

Cerebral palsy

Figure 4.4 An infant with spastic bilateral (quadriplegia) cerebral palsy showing scissoring of the legs from excessive adduction of the hips, pronated forearms and 'fisted' hands.

Summary

Cerebral palsy

- Has many causes. Only about 10% follow hypoxic-ischaemic encephalopathy.
- Usually presents in infancy with abnormal tone and posture, delayed motor milestones and feeding difficulties.
- May be spastic, dyskinetic, ataxic, or a mixed pattern.

Summary

Patterns of cerebral palsy

Type of cerebral palsy	Aetiology	Clinical features
Unilateral cerebral palsy (hemiplegia) Spastic or dystonic	Often due to perinatal middle cerebral artery infarct 	Spastic or dystonic tone, one side of body affected (opposite to the side of the brain lesion) Arm often more affected than leg May have visual field defect on side of hemiplegia Risk of learning difficulties and seizures Often GMFCS level 1 and 2

Bilateral spastic cerebral palsy (diplegia)

Damage to the periventricular areas of developing brain often associated with prematurity.

Leg motor fibres from the homunculus are closest to the ventricles, so legs more affected than arms.

Young child – pattern with walking on their toes with scissoring of the legs.

Older child – crouch gait pattern is typical when the child gets heavier and can't remain on their toes.

Predominantly affects legs. Arms may be subtly affected (supination, fine motor control).
Spasticity is main motor type.
Usually no feeding or communication difficulties and good cognition.
Often associated with squints.
Frequently GMFCS level 1–3

Bilateral spastic cerebral palsy (quadriplegia, 4 limb pattern)	Extensive damage to the periventricular areas of the developing brain, including cortex.		Both arm and leg involvement – predominantly spastic but dystonia often also present. Associated with learning difficulty, feeding difficulties, problems with speech, vision and hearing. Seizures common. At increased risk of hip subluxation and dislocation and scoliosis. Usually dependent on others for activities of daily living Powered mobility a common requirement. Often GMFCS levels 4 and 5.

Dyskinetic cerebral palsy (dystonia, athetosis, chorea)	Perinatal asphyxia – particularly affecting the basal ganglia. Also kernicterus, but this is now rare.	Typical dystonic pattern with open mouth posture and internal rotation and extension of the arms.		Mixture of motor patterns including dystonia, athetosis and chorea. Cognition may be preserved but feeding difficulties are common. Risk of hip deformity and scoliosis. Many are dependent on others for activities of daily living due to their severe movement difficulties even if cognitively normal. Usually GMFCS level 4–5.

Figure 4.5 The different types of cerebral palsy.

Disordered speech and language development

A child may have a deficit in either receptive or expressive speech and language, or both. The deficit may be a delay or a disorder.

Speech and language *delay* may be due to:

- hearing loss
- global developmental delay
- difficulty in speech production from an anatomical deficit, e.g. cleft palate, or oromotor incoordination, e.g. CP
- environmental deprivation/lack of opportunity for social interaction
- normal variant/familial pattern.

Speech and language *disorders* include disorders of:

- language comprehension
- language expression – inability or difficulty in producing speech whilst knowing what is needing to be said
- intelligibility and speech production such as stammering (dysfluency), dysarthria or verbal dyspraxia
- pragmatics (difference between sentence meaning and speaker's meaning), construction of sentences, semantics, grammar
- social/communication skills (autistic spectrum disorder).

Speech and language problems are usually first suspected by parents or primary healthcare professionals. A hearing test and assessment by a speech and language therapist are the initial steps. In early years, there is considerable overlap between language and cognitive (intellectual) development. Involvement of a neurodevelopmental paediatrician and paediatric audiological physician is indicated. Speech and language therapy may be provided on a continuous, burst or review basis. The speech therapist may promote alternative methods of communication such as signing (with Makaton or the Picture Exchange Communication System). Special schooling (usually language units attached to a mainstream primary school) is available but only appropriate for a very few. Many children with early speech and language problems will need learning support at school entry.

There are many tests of language development. These include:

- the Symbolic Toy test, which assesses very early language development
- the Reynell test for receptive and expressive language, used for preschool children.

Abnormal development of social/communication skills (autism spectrum disorders)


Autism Spectrum Disorder
ODD
Specific Learning Difficulties
ADHD
Anxiety
Tourette's
OCD
Developmental Delays
Gifted
Sensory Integration Disorder
Auditory Processing
Depression
ADD

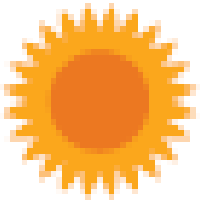
Children who fail to acquire normal social and communication skills may have an autism spectrum disorder. The prevalence of autism spectrum disorder is 3–6 per 1000 live births. The worldwide prevalence is estimated to be 7.6 per 1000 persons. It is more common in boys. Presentation is usually between 2–4 years of age when language and social skills normally rapidly expand. The child presents with a triad of difficulties and associated comorbidities (Box 4.1).

Where only some of the behaviours are present, the child may be described as having autistic features but not the full spectrum.

Asperger syndrome refers to a child with the social impairments of an autism spectrum disorder but at the milder end, and near-normal speech development. Such children still have major difficulties with the give-and-take of ordinary social encounters, a stilted way of speaking and narrow, unusual and often intense interests which they do not share with others, and are often clumsy. In reality, autism spectrum disorders are a continuum of behavioural states ranging from the severe form of autism with or without severe learning difficulties to the milder Asperger syndrome, or to autistic features occurring secondary to other clinical problems.

Autism spectrum disorders are diagnosed by assessing for specific features and seeing if they meet a specific threshold according to the Diagnostic and Statistical manual in the US (DSM5) or International Classification of Diseases. For DSM5 diagnoses, terminologies such as Asperger syndrome and autistic disorder are no longer used, and these are replaced by the term autism spectrum disorder with a description of the particular strengths and difficulties of the child.

Autism is diagnosed by observation of behaviour, including the use of formal standardized tests (Autism Diagnostic Interview, Autism Diagnostic Observation Schedule, and Diagnostic Interview for Social and Communication Disorders). It may arise as the result of different organic processes but in many cases no specific cause can be identified. There is probably multiple aetiology with a genetic component in at least some children. The condition is not the result of emotional trauma or deviant parenting. There is no evidence for a suggested link with the measles, mumps and rubella vaccine.



Autism spectrum disorder:

- Presents at 2–4 years of age with impaired social interaction, speech and language disorder and imposition of routines with ritualistic and repetitive behaviour.
- Usually managed by behaviour modification such as applied behavioural analysis.

Box 4.1 Features of autism spectrum disorders

Impaired social interaction:

- does not seek comfort, share pleasure, form close friendships
- prefers own company, no interest or ability in interacting with peers (play or emotions)
- gaze avoidance
- lack of joint attention
- socially and emotionally inappropriate behaviour
- does not appreciate that others have thoughts and feelings
- lack of appreciation of social cues.

Speech and language disorder:

- delayed development, may be severe
- limited use of gestures and facial expression
- formal pedantic language, monotonous voice
- impaired comprehension with over-literal interpretation of speech
- echoes questions, repeats instructions, refers to self as 'you'
- can have superficially good expressive speech.

Imposition of routines with ritualistic and repetitive behaviour:

- on self and others, with violent temper tantrums if disrupted
- unusual stereotypical movements such as hand flapping and tiptoe gait
- concrete play
- poverty of imagination in play and general activities
- peculiar interests and repetitive adherence
- restriction in behaviour repertoire.

Comorbidities:

- general learning and attention difficulties (about two-thirds)
- seizures (about one-quarter, often not until adolescence)
- affective disorders – anxiety, sleep disturbance
- mental health disorders – attention deficit hyperactivity disorder.

Management

The condition has lifelong consequences of varying degree for the child's social/communication and learning skills. Parents need a great deal of support. They often feel initial guilt that they did not recognize the problem earlier. A wide range of interventions have been promoted over the last 10 years but with little evidence except for applied behavioural analysis, a behaviour modification approach that helps to reduce ritualistic behaviour, develop language, social skills and play, and to generalize use of all these skills. It is currently the most widely accepted treatment approach but requires 25–30 hours of individual therapy each week, so is costly and time consuming. An appropriate educational placement needs to be sought. Some schools incorporate an applied behavioural analysis approach into their teaching methods. Fewer than 10% of children with autism are able to function independently as adults.

Slow acquisition of cognitive skills/general learning difficulty

The term 'learning difficulty or disability' (reflecting cognitive learning difficulties) is now preferred to 'mental retardation' or 'mental handicap'. Medical and educational classification of intelligence quotients (IQs) can be different, with medical models having lower ranges. The educational levels briefed in the following section are useful for general use.

Children with borderline and mild (IQ 70–80) learning difficulties are usually supported by additional helpers (learning support assistants) in mainstream schools, whereas children with moderate (IQ 50–70), severe (IQ 35–50), and profound (IQ < 35) learning difficulties are likely to need the resources of special schools.

Severe or profound learning difficulties are usually apparent from infancy as marked global developmental delay, whereas moderate learning difficulties emerge only as delay in speech and language becomes apparent. Mild learning difficulties may only become apparent when the child starts school or much later.

A child with profound learning difficulties will have no significant language and be completely dependent for all of his/her needs. A child with severe learning difficulties is likely to be able to learn minimal self-care skills and acquire simple speech and language. Both will need high or total supervision and support throughout life.

The prevalence of severe learning difficulty is about 3–4 per 1000 children. Most have an organic cause irrespective of social class, in contrast to moderate learning difficulty (30 per 1000 children) in which children of parents from lower socioeconomic classes are over-represented.

Common causes of developmental delay and learning difficulty are listed in Table 4.2.

Specific learning difficulty

Specific learning difficulty implies that the skill described is more delayed than would be expected for the child's level of cognitive ability. Some examples are described below.

Developmental coordination disorder or dyspraxia

Developmental coordination disorder (developmental dyspraxia) is a disorder of motor planning and/or execution with no significant findings on standard neurological examinations. It is a disorder of the higher cortical processes and there may be associated problems of perception (how the child interprets what he/she sees and hears), use of language and putting thoughts together.

The difficulties may impact on educational progress and self-esteem and suggest the child has greater academic difficulties than may be the case. Features include problems with:

- handwriting, which is typically awkward, messy, slow, irregular and poorly spaced
- dressing (buttons, laces, clothes)
- cutting up food
- poorly established laterality
- copying and drawing
- messy eating from difficulty in coordinating biting, chewing, and swallowing (oromotor dyspraxia). Dribbling of saliva is common.

Assessment and advice are primarily from an occupational therapist and when necessary a speech and language therapist (oromotor skills/speech). A visual assessment may also be helpful. Dyspraxia in its milder form often goes undetected during the first few years of life as the child achieves gross motor milestones at the normal times. With therapy (emphasis on sensory integration, sequencing, executive planning, and where needed speech/language therapy) and maturity, the condition should improve. Verbal dyspraxia is where there are more specific difficulties related to speech production in the absence of muscle or nerve damage. It is considered part of developmental dyspraxia.

Dyslexia

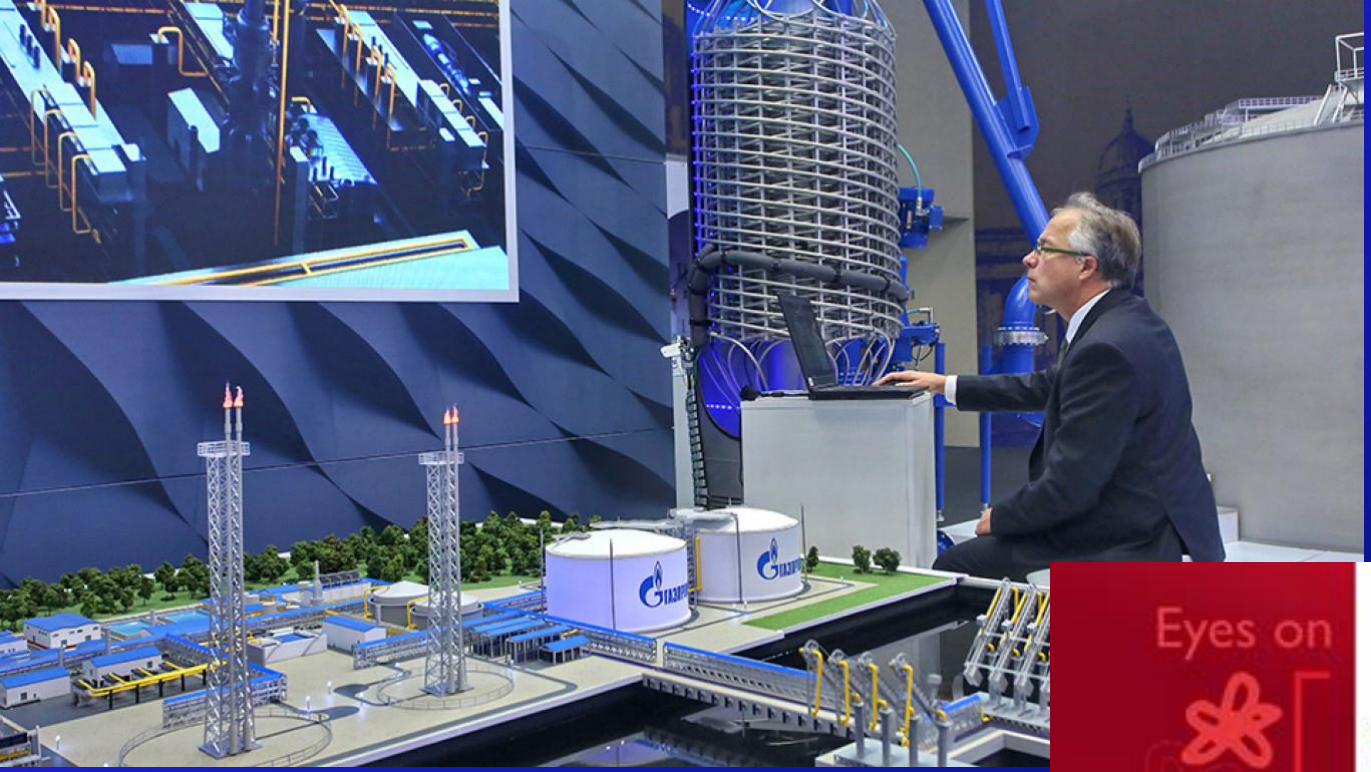
Dyslexia is a disorder of reading skills disproportionate to the child's IQ. The term is often used when the child's reading age is more than 2 years behind his/her chronological age. Assessment needs to include vision and hearing and involves an educational psychologist.

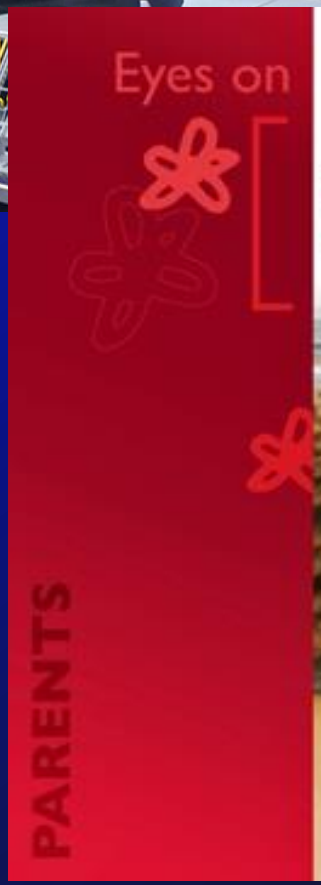
Dyscalculia, dysgraphia

These are disorders in the development of calculation or writing skills.

Disorder of executive functions

Executive functions are a collection of cognitive processes that are responsible for activities such as planning and organization self-regulation, cognitive flexibility, problem solving and abstract reasoning. They are very necessary to function and interact within one's social environment. Deficits in executive function can occur as a consequence of acquired brain injury such as those following hypoxia, infection, stroke or trauma. Executive dysfunction may manifest as poor concentration, forgetfulness, volatile mood, overeating and poor social skills.




Eyes on
PARENTS


Executive functions
Help your child think before acting

Associated comorbidities of specific learning disorders

These are:

- attention deficit disorder
- hyperactivity
- sensory processing disorder (poor sensory integration skills of touch, balance)
- depression, conduct disorders, obsessive compulsive disorder.

Management of specific learning disorders

Assessment may include vision and hearing and assessment by an occupational therapist, physiotherapist, speech and language therapist and educational psychologist. Comorbidities need to be identified. Treatment is aimed at improving skill acquisition, with educational and information technology support as appropriate.

Problems with concentration and attention

Attention deficit hyperactivity disorder

Young children are characteristically lively, some more than others, by virtue of their immaturity. When their level of motor activity exceeds that regarded as normal, they may be termed 'hyperactive' by their parents. This is a judgement that depends upon the parents' standards and expectations. The term can thus incorrectly be used as a complaint about a child who is normally active in overall terms but who can be cheeky and boisterous at times. Such a child is *not* hyperactive, but the parents need advice about how to handle unwanted behaviour.


ATTENTION
UNFOCUSED
DISABILITY
LEARNING
ANGER
ACTIVE
HYPER
BORED
DISTRACTION
LEARNING
BORED
FRUSTRATION
SQUIRM
FIDGET
ATTENTION
ACTIVE
HYPER
UNFOCUSED
ANGER
HYPER CONCENTRATION
ATTENTION
DISTRACTION
ACTIVE
FRUSTRATION
LEARNING
DISABILITY
FRUSTRATION
CONCENTRATION
DISORDER
ANGER
DAYDREAM
DISABILITY
ACTIVE
HYPER
INTERRUPT
ANGER
FRUSTRATION
FIDGET
BORED
Friendship Circle


Attention
Deficit
Hyperactivity
Disorder


ADHD

In the true *hyperkinetic disorder* or *attention deficit hyperactivity disorder* (ADHD), the child is undoubtedly overactive in most situations and has impaired concentration with a short attention span or distractibility. Differences in diagnostic criteria and threshold mean that prevalence rates among prepubertal schoolchildren are variously estimated as between 10–50 per 1000 children, with boys exceeding girls three-fold. There is a powerful genetic predisposition and the underlying problem is a dysfunction of brain neuron circuits that rely on dopamine as a neurotransmitter and which control self-monitoring and self-regulation.

Affected children are unable to sustain attention or persist with tasks. They cannot control their impulses – they manifest disorganized, poorly regulated and excessive activity; have difficulty with taking turns or sharing; are socially disinhibited; and butt into other people's conversations and play. Their inattention and hyperactivity are worst in familiar or uninteresting situations. They also cannot regulate their activity according to the situation – they are fidgety; have excessive movements inappropriate to task completion; lose possessions; and are generally disorganized. Typically, they have short tempers and form poor relationships with other children, who find them exasperating.

The children do poorly in school and lose self-esteem. They may drift into antisocial activities for a variety of reasons, partly because their behaviour drives parents, teachers and peers to use coercion and punishment, which are ineffectual or breed resentment.

In addition to child psychiatric or paediatric evaluation, the child will usually need to be assessed by an educational psychologist.

First-line management in preschool children and school-aged children with mild-to-moderately severe disorder is the active promotion of behavioural and educational progress by offering specific advice to parents and teachers to build concentration skills, encourage quiet self-occupation, increase self-esteem and how to moderate extreme behaviour. Behavioural interventions similar to those embedded in parenting programmes are helpful. These involve having clear rules and expectations and consistent use of rewards to encourage adherence and where appropriate, consequences to discourage unacceptable behaviour.

For those children in whom this is insufficient, hyperactivity responds symptomatically to several types of medication, although this is usually reserved for children older than 6 years of age. Stimulants, such as methylphenidate or dexamphetamine, and non-stimulants, such as atomoxetine, reduce excessive motor activity and improve attention on task and focused behaviour. The usual approach is not to put the child on medication until behavioural and educational progress is actively promoted by the specific measures mentioned earlier. However, in severe cases with high degrees of impairment, simultaneous psychosocial and medical treatment may be required. It may be necessary to continue medication for several years, sometimes into adulthood. Yearly off-medication trial is recommended to evaluate the need for continuing treatment. Specialist supervision is mandatory. Close liaison with the school is required throughout the years of treatment.

The role of diet in the cause and management of hyperactivity is controversial. Current evidence indicates that the sort of diet which aims blindly to reduce sugar, artificial additives or colourants has no effect. A few children display an idiosyncratic behavioural reaction such as excitability or irritability to particular foods. If this seems likely, trying the child on an exclusion of that particular food may be useful. In general, food and drinks with caffeine are not advised. Overzealous dietary exclusion can lead to malnutrition, especially in a child on stimulant medication that may already have the side-effect of appetite reduction.

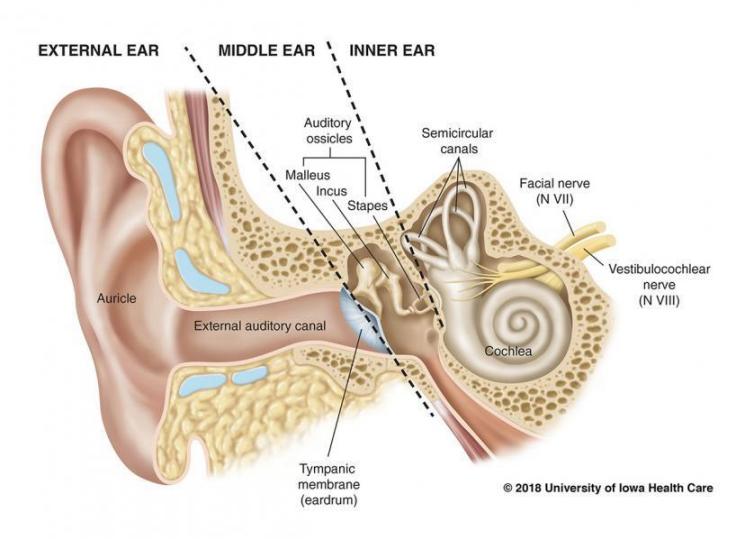
Summary

Attention deficit hyperactivity disorder

- Affects males more than females.
- Clinical features: cannot sustain attention, excessively active, socially disinhibited, easily distracted and impulsive, poor at relationships, prone to temper tantrums, poor school performance.
- Management: educational psychologist assessment, behavioural programmes in school, parenting intervention, medication if necessary.

Hearing impairment

Any concern about hearing impairment should be taken seriously. Any child with delayed language or speech, learning difficulties or behavioural problems should have his/her hearing tested, as a mild hearing



loss may be the underlying cause without parents or other carers realizing. Unilateral hearing loss can cause hearing difficulties when the good ear has an acute ear infection or glue ear. It can also cause difficulty with localizing sounds.

Hearing loss may be:

- sensorineural – caused by a lesion in the cochlea or auditory nerve and is usually present at birth
- conductive – from abnormalities of the ear canal or the middle ear, most often from otitis media with effusion.

The causes, natural history and management of hearing loss are listed in Table 4.5.

Hearing tests are described in Chapter 3. The typical audiogram in sensorineural and conductive hearing loss is shown in Fig. 4.6.

Sensorineural hearing loss

This type of hearing loss is uncommon. In England the incidence of permanent childhood hearing impairment (PCHI) is 0.9 per 1000 live births, with unilateral PCHI adding a further 0.7 per 1000 live births and another 0.7 per 1000 children acquire permanent hearing loss by the age of 10 years. It is usually present at birth or develops in the first few months of life. It is irreversible and can be of any severity, including profound.

The child with severe bilateral sensorineural hearing impairment will need early amplification with hearing aids for optimal speech and language development. Hearing aid use requires close supervision, beginning in the home together with the parents and continuing into school. Children often resist wearing hearing aids because background noise can be amplified unpleasantly. Children with microtia (congenital underdeveloped external ear) and meatal atresia can be helped with bone conduction hearing aids. Cochlear implants may be required where hearing aids give insufficient amplification (Fig. 4.7).

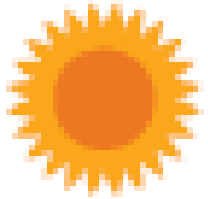
Many children with moderate hearing impairment can be educated within the mainstream school system or in partial hearing units attached to mainstream schools. Children with hearing impairment should be seated in the front of the classroom so that they can readily see the teacher. Gesture, visual context and lip movement will also allow children to develop language concepts. Speech may be delayed, but with appropriate therapy can be of good quality. Modified and simplified signing such as Makaton can be helpful for children who are both hearing-impaired and learning-disabled. Specialist teaching and support service in preschool and school years is provided by peripatetic teachers for children with hearing impairment. Those with profound hearing impairment may need to attend a school for children who are deaf.

Conductive hearing loss

Conductive hearing loss from middle ear disease is usually mild or moderate but may be severe. It is much more common than sensorineural hearing loss. In association with upper respiratory tract infections, many children have episodes of hearing loss, which are usually self-limiting. In some cases of chronic otitis media with effusion, the hearing loss may last many months or years. In most affected children, there are no identifiable risk factors present but children with Down syndrome, cleft palate and atopy are particularly prone to hearing loss from middle ear disease.

Impedance audiometry tests, which measure the air pressure within the middle ear and the compliance of the tympanic membrane, determine if the middle ear is functioning normally. If the condition does not improve spontaneously, medical treatment (decongestant or a long course of antibiotics or treatment of nasal allergy) can be given. If that fails, surgery is considered, with insertion of tympanostomy tubes (grommets) with or without the removal of adenoids. Hearing aids are used in cases where problems recur after surgery.

The decision whether to intervene surgically should be based on the degree of functional disability rather than on absolute hearing loss.



Any child with poor or delayed speech or language must have his/her hearing assessed.

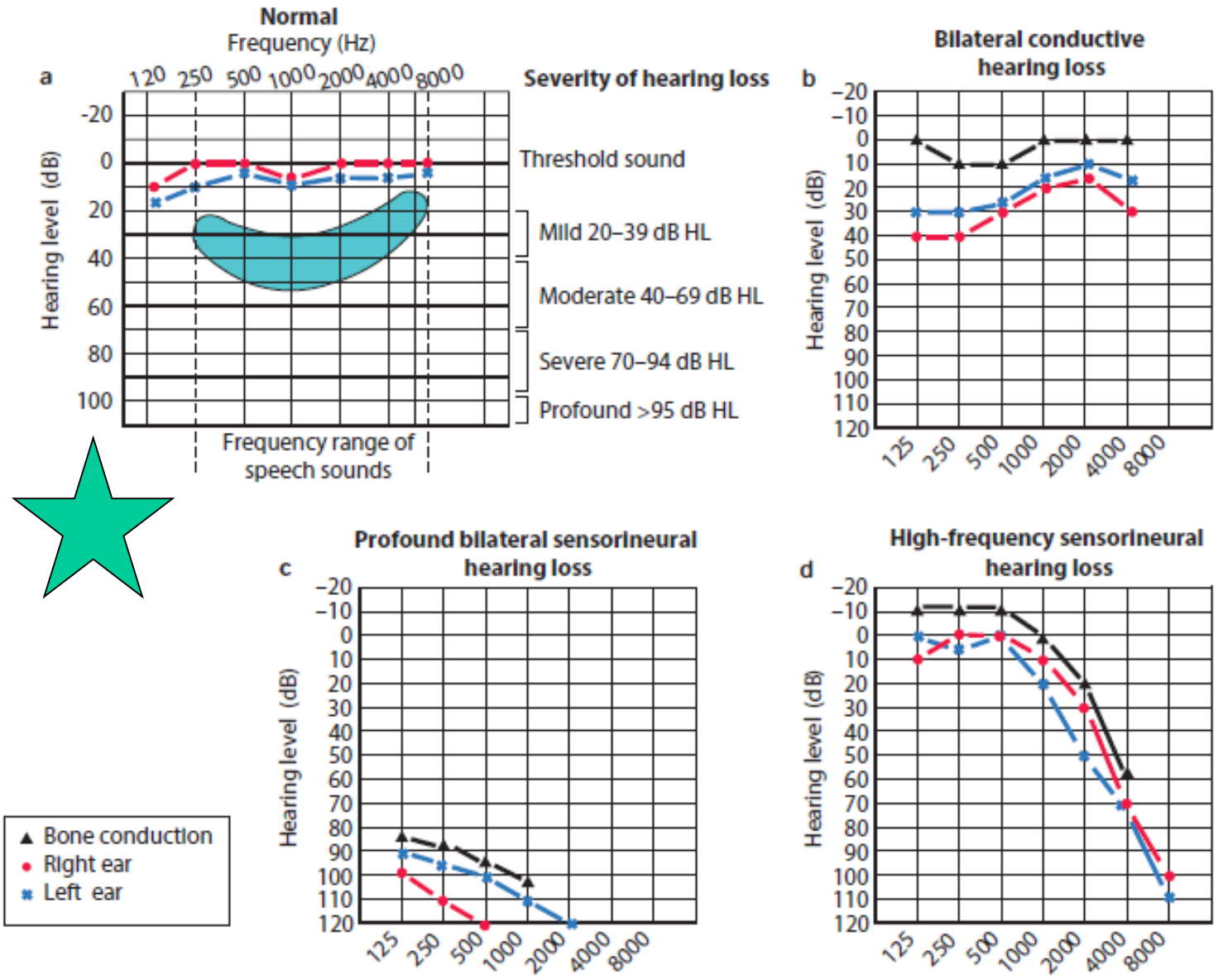
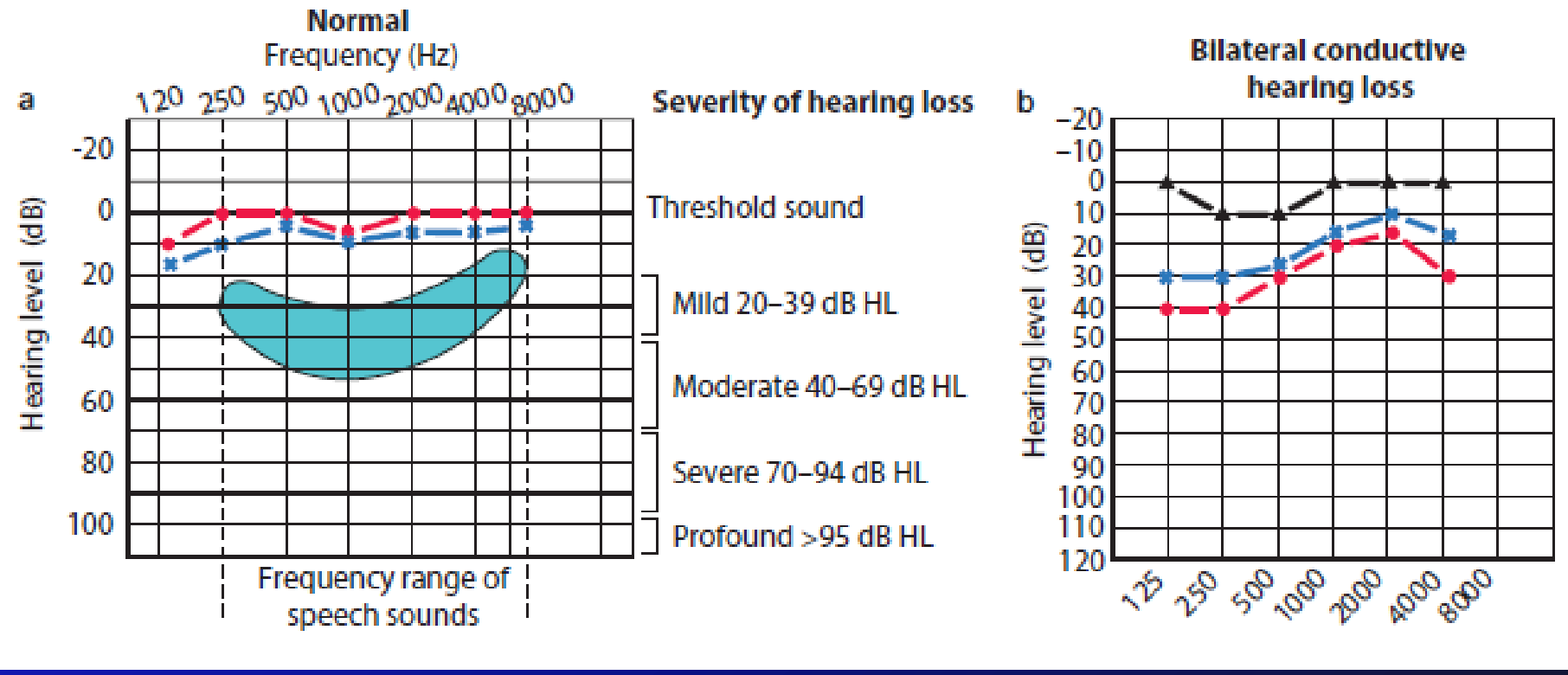
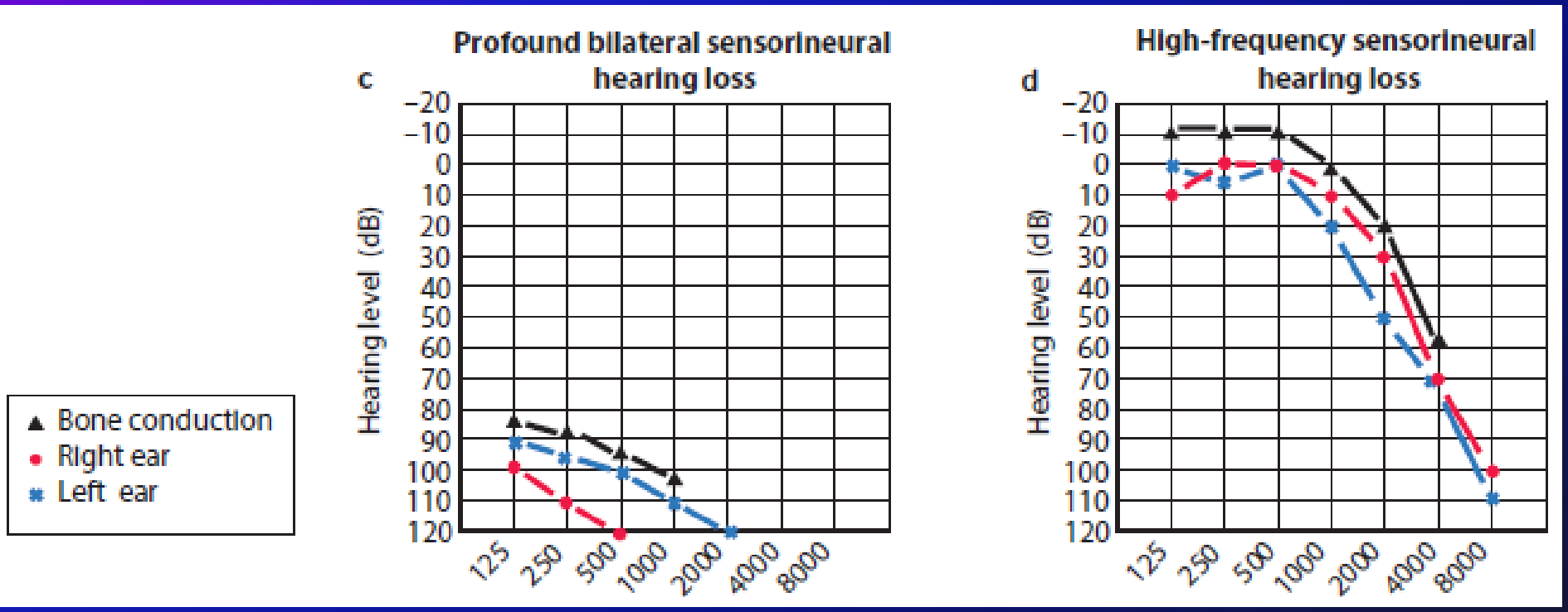


Figure 4.6 (a) Audiogram showing normal hearing and the loudness of normal speech (blue area). The consonants are high-frequency sounds, whereas the vowels are low-frequency sounds; (b) audiogram showing bilateral conductive hearing loss. There is a 30-dB to 40-dB hearing loss in both the right and left ears; (c) audiogram showing bilateral profound sensorineural hearing loss; and (d) audiogram showing bilateral high-frequency sensorineural hearing loss.


Normal
Frequency (Hz)
a
120
250
500
1000
2000
4000
8000
-20
0
20
40
60
80
100
Hearing level (dB)
Severity of hearing loss
Threshold sound
Mild 20–39 dB HL
Moderate 40–69 dB HL
Severe 70–94 dB HL
Profound >95 dB HL
Frequency range of speech sounds
Bilateral conductive hearing loss
b
−20
−10
0
10
20
30
40
50
60
70
80
90
100
110
120
Hearing level (dB)
125
250
500
1000
2000
4000
8000


Bone conduction
Right ear
Left ear
c
Profound bilateral sensorineural hearing loss
Hearing level (dB)
−20
−10
0
10
20
30
40
50
60
70
80
90
100
110
120
125
250
500
1000
2000
4000
8000
d
High-frequency sensorineural hearing loss
Hearing level (dB)
−20
−10
0
10
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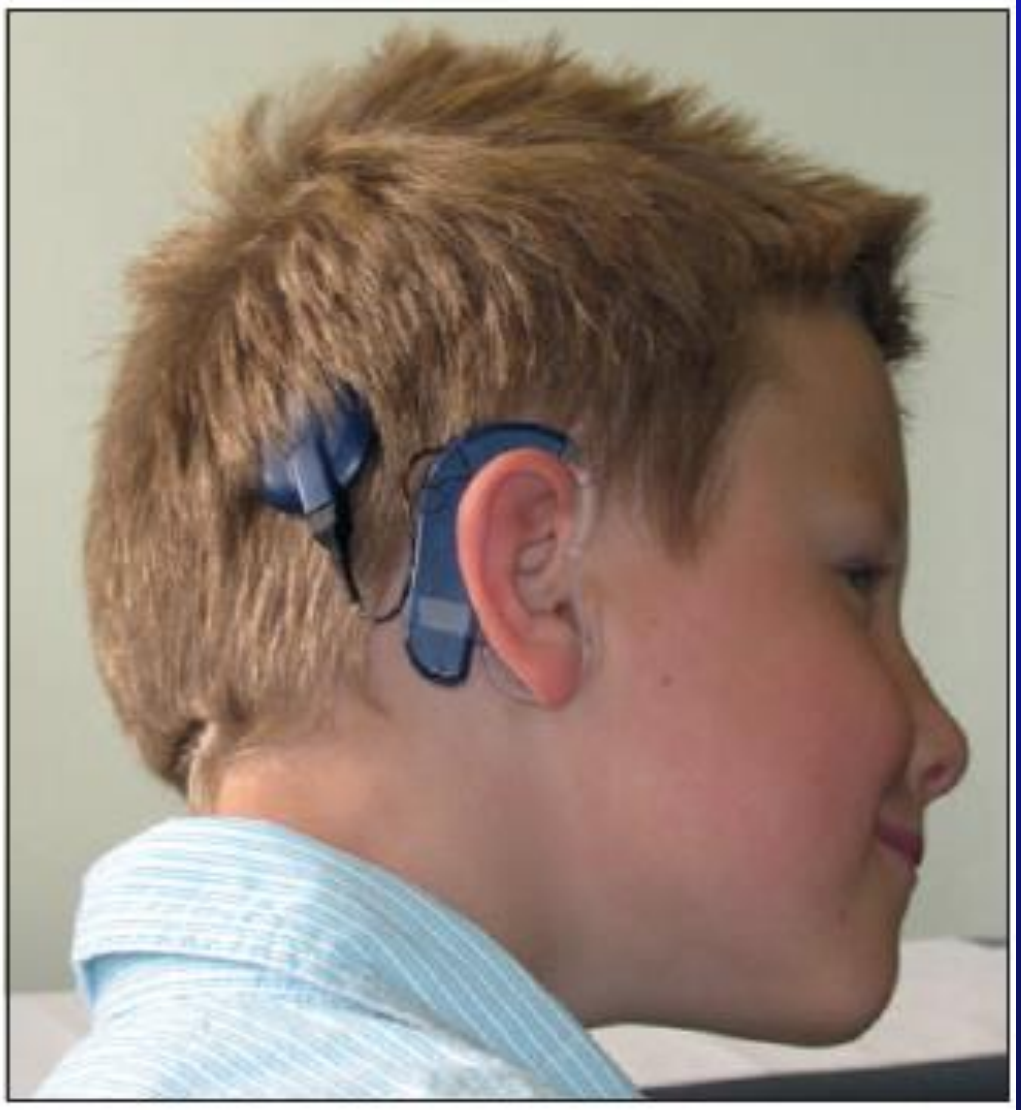


Figure 4.7 Cochlear implant. There is a microphone to detect sound, a speech processor and a transmitter and receiver/stimulator. They convert speech into electric impulses, which are conveyed to the auditory nerve, bypassing the ear. It provides a deaf person with a representation of sounds.

Table 4.5 Causes and management of hearing loss

	Sensorineural	Conductive
Causes	Genetic (the majority) Antenatal and perinatal: • congenital infection • preterm • hypoxic-ischaemic encephalopathy • hyperbilirubinemia Postnatal: • meningitis/encephalitis • head injury • drugs, e.g. aminoglycosides, furosemide (frusemide) • neurodegenerative disorders	Otitis media with effusion (glue ear) Eustachian tube dysfunction: • Down syndrome • cleft palate • Pierre Robin sequence • midfacial hypoplasia Wax (only rarely a cause of hearing loss)
Hearing loss	May be profound (>95-dB hearing loss)	Maximum of 60-dB hearing loss
Natural history	Does not improve and may progress	Intermittent or resolves
Management	Amplification or cochlear implant if necessary	Conservative, amplification or surgery

Summary

Hearing loss

Sensorineural hearing loss:

- is usually present at birth and is irreversible
- early amplification with hearing aids or cochlear implants is needed for severe hearing impairment for optimal speech and language development
- assistance is required from peripatetic teachers for children with moderate/severe hearing impairment.

Conductive hearing loss:

- is usually due to middle ear disease, often otitis media with effusion
- is usually mild or moderate and transient
- consider insertion of tympanostomy tubes (grommets) with or without the removal of adenoids if it does not resolve.

Abnormalities of vision and the ocular system

Normal visual development and tests of vision are described in Chapter 3.

Visual impairment may present in an infant or young child with:

- obvious ocular malformation (e.g. anophthalmia)
  - absent red reflex or white reflex (leukocoria), which may be due to opacification of intraocular structures, corneal abnormalities or intraocular tumour (retinoblastoma)
- not smiling responsively by 6 weeks' post-term

- concerns about poor visual responses, including poor eye contact
- roving eye movements
- nystagmus
- squint.

Any infant presenting with an ocular abnormality needs prompt referral to an ophthalmologist as some underlying conditions are sight threatening, and retinoblastoma is life threatening.

Nystagmus

This is a repetitive, involuntary, rhythmical eye movement. It is usually horizontal but can be vertical. It may be found in association with a structural eye problem (sensory defect nystagmus), but can also be a consequence of a problem at the cortical level. Nystagmus which is a manifestation of an eye problem, may improve over time. If no structural eye (or brain) problem is found, a diagnosis of idiopathic nystagmus is made.

Squint (strabismus)

In this common condition there is misalignment of the visual axes. Squint should be assessed in order for the underlying cause to be identified and treated where possible. There may be a family history. Transient misalignment is common up to 3 months of age. Marked epicanthic folds may give an appearance of a squint. Any infant with a squint should have red reflexes checked. Squints persisting beyond 3 months of age should be referred for a specialist ophthalmological opinion. The most common underlying cause is refractive error, but cataracts, retinoblastoma, and other intraocular causes must be excluded.

Squints are commonly divided into:

- *concomitant* (non-paralytic, common) – usually due to a refractive error in one or both eyes. Correction of the refractive error with glasses often corrects the squint. The squinting eye most often turns inwards (convergent), but there can be outward (divergent) or, rarely, vertical deviation
- *paralytic* (rare) – varies with gaze direction due to paralysis of the motor nerves. This can be sinister because of the possibility of an underlying space-occupying lesion such as a brain tumour.

Corneal light reflex test

Non-specialists can use this test to detect squints (Fig. 4.8). A pen torch is held at a distance to produce reflections on both corneas simultaneously. If the light reflection does not appear in the same position in the two pupils, a squint is present. However, a minor squint may be difficult to detect.

Cover test

The child is encouraged to look at a toy/light. If the fixing eye is covered, the squinting eye will move to take up fixation. (Fig. 4.9). The test should be performed with near (33 cm) and distant (at least 6 m) objects, as certain squints are present only at one distance. These tests are difficult to perform and reliable results are best obtained by an orthoptist or ophthalmologist.

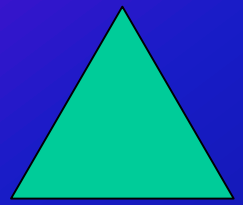
Refractive errors

Hypermetropia (long sight)

This is the most common refractive error in young children. Mild hypermetropia is common in early childhood and is overcome through the process of accommodation – changing the shape of the lens in the eye. Hypermetropia can be corrected with convex (plus) lenses. These make the eye look bigger. Mild hypermetropia may not need spectacle correction.

Myopia (short sight)

This is relatively uncommon in young children, presenting usually in adolescence. However, in children born preterm it is the most common refractive error and may present at a younger age. Myopia can be corrected with concave (minus) lenses. These make the eye look smaller.



Astigmatism (abnormal corneal curvature)

Minor degrees of astigmatism are common and may not cause problems or require correction. Unilateral astigmatism can result in amblyopia.

Amblyopia

This is a potentially permanent reduction of visual acuity in an eye that has not received a clear image. It affects 2–3% of children. It is usually unilateral but can be bilateral. The most common causes of amblyopia are squint, refractive errors and obstruction to the visual pathway, e.g. cataract. Amblyopia may occur in squint when the brain is unable to combine the markedly differing images from each eye – the vision from the squinting eye is 'switched off' to avoid double vision. Treatment entails tackling the underlying condition, together with patching of the 'good' eye for specific periods of the day to force the 'lazy' eye to work, and therefore develop better vision. Early treatment is essential, as after 7 years of age improvement is unlikely. Considerable encouragement and support should to be given often to both the child and parents, as young children usually dislike having their better eye patched. Amblyopia may be asymptomatic, and is the main target condition for preschool vision screening in the UK.

Severe visual impairment

This affects 1 in 1000 live births in the UK, bu[illegible]her in developing countries. The main causes are listed in Box 4.2. Recent epidemiological studies suggest that in the UK up to 50% of children have cerebral pathology as the underlying cause; about one-third of cases are hereditary, affecting eye structures. In developing countries, acquired causes such as infection are more prevalent.

Investigations may include an electroretinogram or visual evoked potentials. When visual impairment is of cortical origin, resulting from cerebral damage, examination of the eye, including the pupillary responses, may be normal.

Although few causes of severe visual impairment can be cured, early detection allows certain elements to be treated and timely advice can be given on supporting developmental progress. In the UK, this advice is usually provided by peripatetic teachers for children with visual impairment, who work with families from the time of diagnosis, irrespective of the child's age. Input from a paediatrician and other members of the child development team are also required. Partially sighted children may benefit from provision of low vision aids, high-powered magnifiers and small telescopic devices and computers. Although many severely visually impaired children have a visual disability alone, at least half have additional neurodevelopmental problems.

Squints

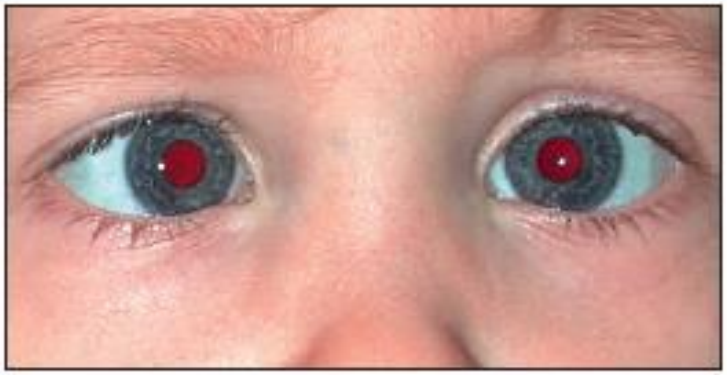


Figure 4.8 Corneal light reflex (reflection) test to detect a squint. The reflection is in a different position in the two eyes because of a small convergent squint of the right eye.

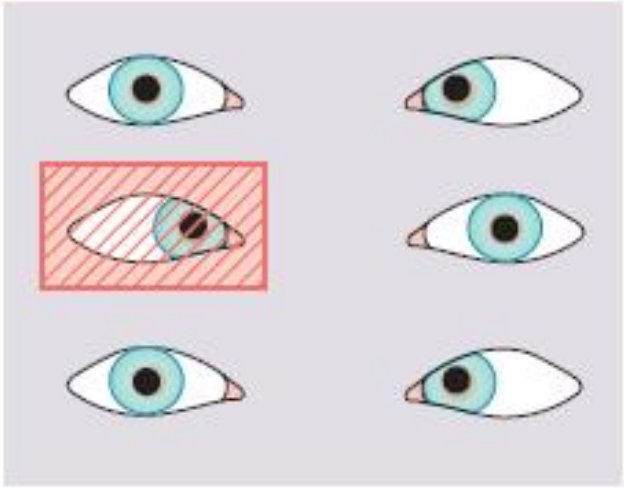


Figure 4.9 The cover test is used to identify a squint. If the fixing eye is covered, the squinting eye moves to take up fixation. This diagram shows a left convergent squint.

Box 4.2 Causes of visual impairment

Genetic	Antenatal and perinatal	Postnatal
Cataract	Congenital infection	Trauma
Albinism	Retinopathy of prematurity	Infection
Retinal dystrophy	Hypoxic-ischaemic encephalopathy	Juvenile idiopathic arthritis
Retinoblastoma	Cerebral abnormality/damage	
	Optic nerve hypoplasia	

Summary

Regarding vision

- Visual impairment and ocular abnormalities including refractive errors are more common in children with neurodevelopmental problems.
- An absent red reflex or white reflex or a squint persisting after 3 months of age – refer to an ophthalmological opinion.
- Abnormal eye movements in an infant, absence of responsive smiling, or parental concern about vision at any age – consider visual impairment and refer to an ophthalmological opinion.
- Testing for squints – corneal light reflex (reflection) test for the non-specialist, cover test for the specialist.
- Amblyopia may be asymptomatic; treatment often includes patching of the 'good' eye for short periods each day.

Multidisciplinary child development services

Although children with a wide range of conditions have additional needs, the term 'special needs' is usually used for children with developmental problems and disabilities. In order to optimize the child's assessment and care on an ongoing basis, child development services in the UK are based on geographic areas and are secondary care services.

A child development service:

- is multidisciplinary with predominantly health professionals (paediatrician, physiotherapist, occupational therapist, speech and language therapist, clinical psychologist, specialist health visitor, dietician) in the team but often also includes a social worker (Fig. 4.10)
- is multiagency (Fig. 4.11) and may include health, social services, education, volunteers, voluntary agencies, parent support groups
- aims to provide a coordinated service with good inter-agency liaison to meet the functional needs of the child and optimize his/her care
- may provide multidisciplinary support and monitor children up to school-leaving age (16–19 years)
- maintains a register of children with disabilities and special needs
- has emphasis on children's needs within the community (home, nursery, school), regardless of its location.

Child development services in the UK now usually use the Common Assessment Framework to allow multi-disciplinary sharing of information. The emphasis is on:

- diagnosis
- assessment of functional skills: mobility, hand function, vision, hearing, communication, behaviour, social/self-care skills and learning
- provision of therapy
- regular review
- a coordinated approach to care (multidisciplinary, multiagency).

Many children with special needs have medical problems which require investigation, treatment, and review. Good inter-professional communication is vital for well-coordinated care. This is assisted by all professionals keeping entries in the child's personal child health record up to date.

In addition to locally organized child development services, specialist neurodisability services are required for:

- rehabilitation following acquired brain injury
- surgery for cerebral palsy, scoliosis
- gait analysis
- spasticity management, including botulinum toxin injections to muscles
- epilepsy unresponsive to two or more anticonvulsants or where there is severe cognitive and behavioural regression related to epilepsy
- complex communication disorders – diagnosis and therapeutic intervention
- mixed complex learning problems, often with neuropsychiatric comorbid symptoms
- provision of communication aids (Fig. 4.12)
- sensory impairments (e.g. cochlear implants)

- children with severe visual and hearing impairment
- specialized seating/wheelchairs and orthoses (Fig. 4.13)
- management of movement disorders (e.g. continuous infusion of intrathecal baclofen, selective dorsal rhizotomy and deep brain stimulation to basal ganglia).

Needs are likely to change over time, with key stages being at transition to school and adult services. A care plan should be developed at each stage; it may include education and social care as well as health. Care plans should be shared with the child and family and then regularly reviewed. Involvement with specialist services may be of variable frequency throughout childhood. Collaboration across services is vital in promoting a service tailored around the child and family.

Summary

Children with developmental problems and disabilities

- Are looked after by local multidisciplinary child development services.
- Often have complex medical needs.
- Need regular review, as needs change with time, as will the child's ability to participate in it.
- Require coordination of care between the family and the many professionals involved, as well as close liaison with education and social services.

Hearing
Conductive or sensorineural hearing impairment

Vision
Squint
Impaired visual acuity
Visual field deficits

Orthopaedic
Hip subluxation/dislocation
Fixed joint contractures
Dynamic muscle contractures
Painful muscle spasm
Spinal deformity
Osteoporosis/fractures

Specialist health visitor
Helps coordinate multidisciplinary and multi-agency care
Advice on development of play or local authority schemes e.g. Portage

Dietician
Advice on feeding and nutrition

Social worker/Social services
Advice on benefits: disability, mobility, housing, respite care, voluntary support agencies
Day nursery placements
Advocate for child and family
Register of children with special needs

Psychologist (clinical and educational)
Cognitive testing
Behaviour management
Educational advice

Gastrointestinal
Gastro-oesophageal reflux
Oromotor incoordination
Aspiration of food or saliva
Constipation

Urogenital
Urinary tract infection
Delay in establishing continence
Unstable bladder
Vesico-ureteric reflux
Neuropathic bowel and bladder

Common medical problems

Child Development Services

Paediatrician
Assessment, investigation and diagnosis
Continuing medical management
Coordination of input from therapists and other agencies – health, social services, education

Respiratory
Respiratory infections
Aspiration pneumonia
Chronic lung disease
Sleep apnoea

Neurological
Epilepsy
Microcephaly/hydrocephalus
Cerebral palsy

Nutrition
Poor weight gain
Faltering growth

Behaviour
Organic or reactive
Sibling behaviour
Parental distress

Speech and language therapist
Feeding
Language development
Speech development
Augmentative and alternative communication (ACC) aids e.g. Makaton sign language, Bliss symbol boards, voice synthesisers

Occupational therapist
Eye–hand coordination
Activities of daily living (ADL) – feeding, washing, toileting, dressing, writing
Seating
Housing adaptations

Physiotherapist
Balance and mobility
Postural maintenance
Prevention of joint contractures, spinal deformity
Mobility aids, orthoses

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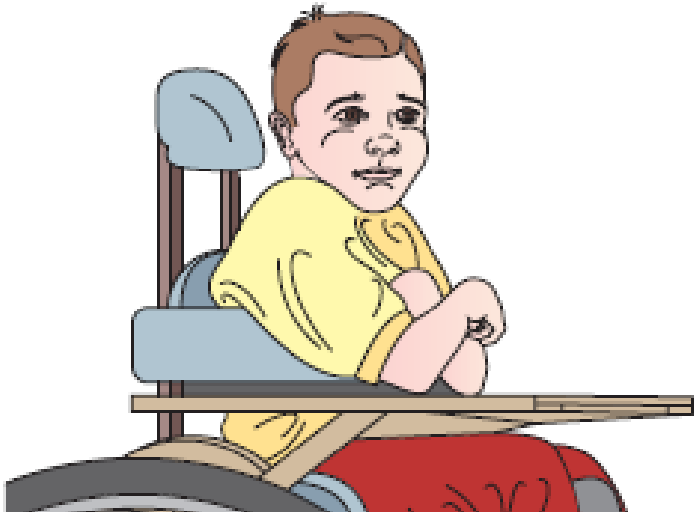

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igure 4.10 Common medical conditions and the many professionals in the child development services nvolved in the care of children with developmental problems.

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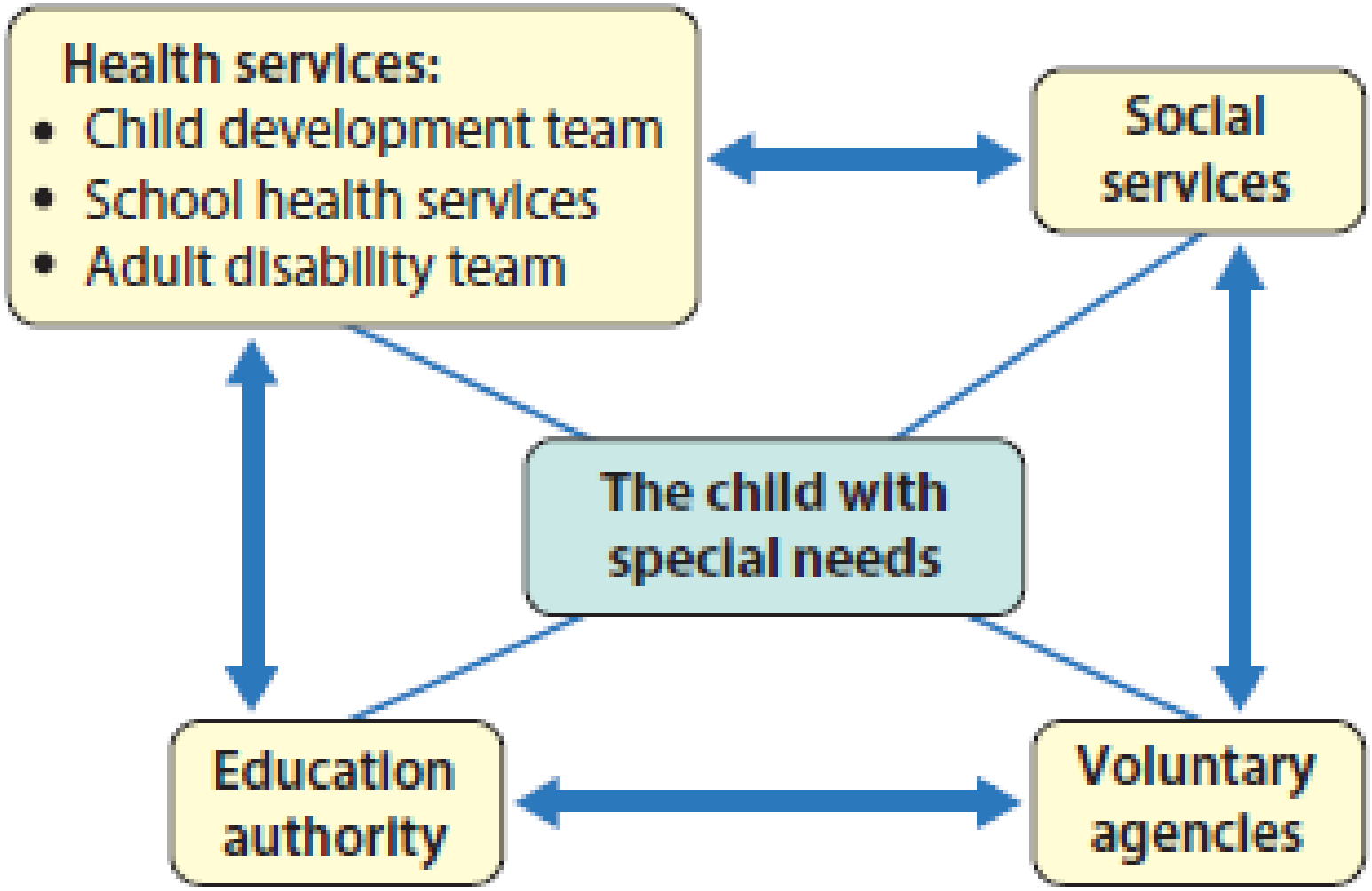


Figure 4.11 Children with special needs are supported by the integrated inputs of health and social services, local education authorities and voluntary agencies.

Figure 4.12 An example of a touchscreen speaking communication aid to assist children who may have speaking and movement difficulties.

a

b

Figure 4.13 (a) A boy with spastic cerebral palsy is able to walk with the help of a frame; and (b) a motorized wheelchair that enables this young person with cerebral palsy to be mobile.

Education

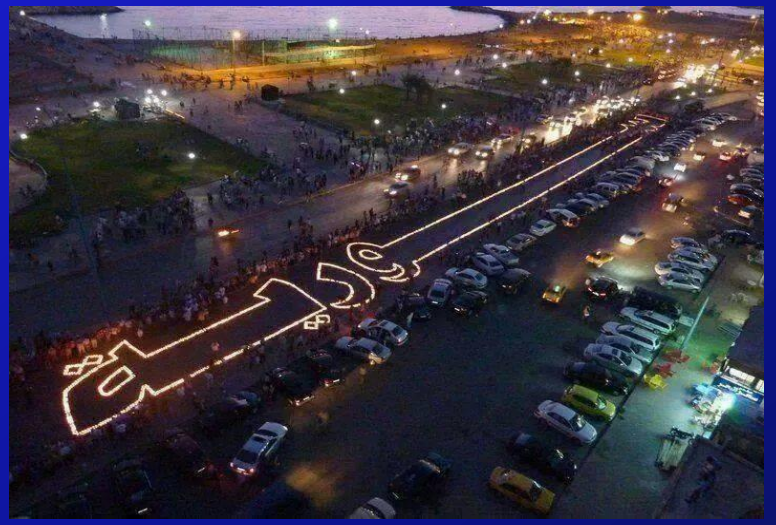
Children with special educational needs should receive educational input according to their requirements, including integration into mainstream schooling whenever possible. However, there are special needs' schools for children with more significant learning and/or physical disabilities. There are also specialist educational placements for children with certain conditions such as severe visual or hearing impairment or autism spectrum disorders. These specialist educational settings have access to a higher level of therapies (physiotherapy, occupational therapy and speech and language therapy) and specialist teaching than are available through a mainstream educational placement. Assessment and support for behavioural and learning needs may come from an educational or clinical psychologist.

Transition of care to adult services

In the UK, adult disability services are, in general, still poorly developed by comparison with those provided for children. Young adults with severe learning and physical disabilities are supported by Adult Learning Disability Teams, but there is only limited national provision for those with mild or moderate learning disabilities or with a predominantly physical disability. Major problems for young adults with disabilities include social issues around care, housing, mobility, finance, leisure, employment, and genetic and sexual counselling. Health information must be properly transferred from child to adult health services if reinvestigation of already well-clarified conditions is to be avoided.

The rights of disabled children

Irrespective of their disability, the aspirations and rights of children, as affirmed by the United Nations Convention on the Rights of the Child, need to be respected. Technological advances to improve mobility, communication and emotional expression are helping enable people with disability to better achieve their full potential, rather than being held back by their disability. However, this requires skilled assistance and adequate resources. Prominent public figures who function effectively despite disabilities help to make the public appreciate what can be achieved and serve as an inspiration to those with disabilities. The World Health Organization stresses the important outcomes of activity and participation. Any health interventions for people with disability either on an individual level or in society as a whole should aim to also improve these outcomes.



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