CASE SIX

Short case number: 3_12_06
Category: Children & Young People
Discipline: Paediatrics Medicine
Setting: Paediatric Outpatients
Topic: Developmental delay_cerebral palsy.

Case

Henry Jacobsen is 18 months old; he has been referred to the paediatric outpatients by his GP for assessment of his developmental delay. Henry's parents are concerned that he is not yet walking and is unsteady when he tries to stand on his own. On reviewing the referral letter you also note that the GP has commented that the muscular tone in Henry's lower limbs is increased and the GP is questioning whether he has cerebral palsy.

Questions

1. In considering the possible causes of cerebral palsy what key questions would you ask as part of your medical history?
2. What examination findings would support a diagnosis of cerebral palsy?
3. What associated disorders would you assess for as part of your physical examination?
4. Henry's parents ask you what cerebral palsy is; what would you explain to them?
5. Briefly outline the classification system for cerebral palsy.
6. Children with cerebral palsy often require multidisciplinary management depending on the associated disabilities and health problems. In a table summarise the associated disabilities and health problems that can occur in conjunction with cerebral palsy, outlining the management of these problems and the role of the health and allied health care team in the management.

Suggested reading:

- South M, Isaacs D editors. Practical Paediatrics. 7th edition. Edinburgh: Churchill Livingstone; 2012.

ANSWERS

1 . Cerebral palsy is not a single disorder but a group of disorders with diverse implications for children and their families. It is the most common physical disability in childhood. The cause of cerebral palsy is unknown in many children.

It is helpful to consider the timing of the brain insult:
- prenatal events are thought to be responsible for approximately 75% of all cases.
- perinatal events contribute 10-15%.
- postnatal causes account for about 10% of all cases.

As part of the medical history, Henry's mother needs to be asked the following questions:
- is there a family history of cerebral palsy or any other developmental delay
- were there any abnormalities detected with Henry on routine imaging during the pregnancy and if so were there other forms of imaging conducted (e.g. MRI)
- were there any maternal infections during the first and second trimester, for instance, the TORCH group of organisms (toxoplasmosis, rubella, cytomegalovirus and herpes simplex virus), may cause cerebral palsy
- is there a history of thyroid disease, is there a family history of thyroid disease to suggest a possible iodine deficiency.
- was Henry premature and if so how early (the rate of cerebral palsy in children born before 33 weeks is up to 30 times higher than in those born at term. Periventicular leukomalacia is a common radiological finding in premature children with cerebral palsy. It is caused by an ischaemic process in the watershed zone that exists in the periventricular white matter of the immature brain).
- were there any problems associated with either labour or the delivery e.g. prolonged, obstructed labour, antepartum haemorrhage or cord prolapse
- was Henry of low birth weight
- was Henry the product of a multiple pregnancy and if so did the other sibling(s) survive
- did Henry have any problems
- have there been any major illnesses since birth e.g. meningitis, septicaemia
- have there been any injuries since birth

2. The initial presentation of cerebral palsy (CP) includes early hypotonia, followed by spasticity. Generally, spasticity does not manifest until at least 6 months to 1 year of age.
Observation may reveal abnormal neck or truncal tone (decreased or increased, depending on the age and the type of CP); asymmetric posture, strength or gait, or abnormal co-ordination.
Patients with CP may show increased reflexes, indicating the presence of an upper motor neurone lesion. They may also present with the persistence of primitive reflexes, such as the Moro (startle reflex) and asymmetric tonic neck reflexes. CP may also include the underdevelopment or absence of postural or protective reflexes (extending one arm when sitting up).
Patients with spastic CP exhibit spasticity (a velocity-dependent increase in tone).
It may be evident by a tendency to keep the elbow in a flexed position or the hips flexed and adducted with the knees flexed and valgus and the ankles in equinus, resulting in toe-walking.

Patients with dyskinetic or extrapyramidal CP may have decreased head and truncal tone and defects in postural control and motor dysfunction such as the following: - athetosis (slow, writhing, involuntary movements, particularly in the distal extremities)

- chorea (abrupt, irregular, jerky movements) or choreoathetosis
- dystonia (slow rhythmic movements with increased muscle tone and abnormal postures e.g. in the jaw and upper extremities)

Classic physical presentations of the different types of CP include the following:

- Spastic hemiplegic CP
  - · one-sided upper motor neurone deficit
  - · arm generally affected more than leg, relative weakness on one side, gait may be characterized by circumduction of the lower extremity on the affected side.
  - · specific learning disabilities
  - · oromotor dysfunction
  - · possible unilateral sensory defects
  - · visual field deficits (homonymous hemianopsia) and strabismus
  - · seizures
- Spastic diplegic CP
  - · upper motor neurone findings in the legs more than the arms
  - · scissoring gait pattern with hips flexed and adducted, knees flexed and valgus and ankles in equinus, resulting in toe-walking
  - · learning disabilities and seizures less commonly than in spastic hemiplegia
- Spastic quadriplegic CP
  - · all limbs affected, either full-body hypertonia or truncal hypotonia with extremity hypertonia
  - · oromotor dysfunction
  - · increased risk of cognitive difficulties
  - · multiple medical complications
  - · seizures
  - · legs generally affected equally or more than arms
  - · categorized as double hemiplegic if arms more involved than legs
- Dyskinetic (extrapyramidal) CP
  - · early hypotonia with movement disorder emerging at age 1- 3 years
  - · arms more affected than legs
  - · deep tendon reflexes usually normal to slightly increased
  - · some spasticity
  - · oromotor dysfunction
  - · gait difficulties
  - · truncal instability
  - · risk of deafness in those affected by kernicterus.

3. Associated disorders
   - visual problems occur in about 40% of children with CP and include strabismus, refractive errors, visual field defects and cortical visual impairment
   - hearing deficits occur in 3-10% of children with CP. High-frequency hearing loss may be found in children with congenital rubella or other viral syndromes.
   - speech and language problems: receptive and expressive language delays and articulation problems occur.
   - epilepsy occurs in up to 50% of children with CP, most commonly in those with

severe motor problems.

- cognitive impairments: while intellectual disabilities and learning problems are common, there is a wide range of intellectual ability in children with CP and children with severe physical disabilities may have normal intelligence. Perceptual difficulties are also frequent.

Some children with cerebral palsy have only a motor disorder.

4. The term cerebral palsy loosely translates to 'brain paralysis'. It is not a single illness but an 'umbrella' termto describe a group pf permanent disorders of movement and posture, causing activity limitation, that are attributed to a non-progressive insult that occurred in the developing foetal or infant brain (normally before the age of 2 years). The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, behaviour, epilepsy and by secondary musculoskeltal problems
   There are many different causes and a wide range in which the movement disorder presents. It is the most common physical disability in childhood, in developed countries the chance of it occurring being between 2.0 and 2.5 cases per 1000 live births. A number of factors increase this risk however the cause is unknown in many children. To attempt to identify a possible cause appropriate imaging and also blood tests may need to be undertaken.
   Patients with milder forms of cerebral palsy have a life expectancy close to the general population although still somewhat reduced.

5. There are three major ways in which cerebral palsy is classified – by type, by topographical distribution and by the severity of the motor disorder.
   Type of motor disorder
   Cerebral palsy is a disorder of movement (difficulties with voluntary movement and/or abnormal movements), posture and muscle tone. Children with cerebral palsy may present with various types of movement disorder.
   Spastic cerebral palsy (70%)
   This is the most common type. Spasticity involves increased muscle tone with characteristic clasp knife quality. Children with spasticity often have underlying weakness. In spastic cerebral palsy, there is damage to the motor cortex or corticospinal tracts, in contrast to dyskinetic and ataxic cerebral palsy, which are associated with abnormalities of the basal ganglia and cerebellum, respectively.
   Dyskinetic cerebral palsy (10-15%)
   This refers to a group of cerebral palsies with involuntary movements and is characterized by abnormalities of tone involving the whole body. Several terms are used within this group:
   - dystonia is a syndrome of sustained muscle contractions, frequently causing twisting and repetitive movements of abnormal postures.
   - athetosis refers to slow writhing movements involving the distal parts of the limbs.
   - chorea is the term for rapid jerky movements.

   Ataxic cerebral palsy (less than 5%)
   Children have a fine tremor, more noticeable when movements are initiated, as well as poor balance and hypotonia. Ataxia is associated with other neurological conditions that must be excluded before this diagnosis is made. Some children have a mixed motor disorder.
   The topographical distribution.

The terms diplegia, hemiplegia and quadriplegia are used and generally apply to children with spastic cerebral palsy as the other types usually involve four limbs:

· the term *diplegia* is used where the predominant problem is in the lower limbs. There is usually some upper limb involvement, which may be subtle. The majority of these children have normal intelligence. Spastic diplegia is the pattern most commonly seen in premature infants who have the radiological finding of periventricular leukomalacia.

· children with spastic *hemiplegia* usually have normal intelligence, frequently have epilepsy (50-70%), may have sensory impairments in the upper limb and may have visual deficits (homonymous hemianopsia)

· children with spastic *quadriplegia* frequently have problems such as intellectual disability, epilepsy and visual difficulties. There is often poor trunk control and oromotor difficulties in addition to four limb involvement.

Severity of the motor disorder

The gross motor function classification system (GMFCS), provides information about the movement problems of children with cerebral palsy based on their motor abilities and their need for walking frames, wheelchairs and other mobility devices. There are five levels: children in levels I and II walk independently, children in level III generally need walking frames or elbow crutches and children in levels IV and V use wheelchairs. This classification system does not consider cognitive and other deficits which may have a profound effect on the eventual outcome.

Using the GMFCS, growth motor development curves have been constructed that provide some guide to prognosis for motor development. There are now classification systems for the use of your hands (Manual Ability Classification - MACS) and communication (communication function classification system (CFCS).

6.

Associated disabilities and health problems	**Management**	**Role of the health and allied health care team**
Hearing deficits	Tailored to individual child, e.g. hearing aids etc.	Assessment and monitoring by audiologist
Visual problems	Correction of strabismus and refractive errors, assessment of visual field defects and cortical visual impairment	Assessment and monitoring by Optometrist and Paediatric Ophthalmologist
Failure to thrive, under nutrition, obesity, constipation	Dietary advice sought to determine adequacy of nutrient and calorie intake. Management of nasogastric or gastroscopy feeds. Dietary advice for bowel regularity	Nutritionist /dietician
Abnormal muscle tone, impaired movement skills and postural difficulties	Practical advice to parents on positioning, handling and play to minimize motor problems Post-operative rehabilitation phase following orthopaedic procedures. Botolinum toxin injections and anti-spasticity medications	Physiotherapist Trained massage therapists with special interest and expertise Rehabilitation specialist will administer botox injections
Impaired functional skills causing restriction in activity levels	Assist parents to develop their child's upper limb and self-care skills, and also recommend suitable toys, equipment and home adaptations	Occupational therapists

Associated disabilities and health problems	Management	Role of the health and allied health care team
Impaired communication skills	Assist in the development of communication skills, including advising about augmentative communication systems for children with limited verbal skills. They provide guidance about feeding difficulties and saliva control problems.	Speech pathologists
Dental problems	Monitor and treat dental problems as required	Dentist
Orthopaedic problems e.g. contractures, hip subluxation or dislocation, knee flexion contractures, equinus deformity at the ankle	Hip X-rays at yearly intervals. Flexion knee contractures may require hamstring surgery. Conservative treatment of equinus with orthoses, inhibitory casts and botulinum toxin therapy or surgery in older children. 'Single event multilevel surgery' with a single hospitalization for surgery at several different levels, the aims are to correct deformities and to improve both the appearance and efficiency of walking	Paediatric orthopaedic surgeon Physiotherapist Rehabilitation specialist
Seizures , muscle contractions, gastro-oesophageal reflux disease (GORD), chronic lung disease due to aspiration from oromotor dysfunction or severe GORD, osteoporosis, hydrocephalus	Oral medications to manage seizures, GORD, lung infections etc. Use of Botulinum toxin for control of increased tone and spasticity to aid ambulation	Paediatrician, paediatric Specialists - Neurologist, Neurosurgeon,Gastroenterologist , Endocrinologist,
Ascent of testes with time (secondary to chronic spasm of the cremasteric muscle)	Scrotal orchidopexy	Paediatric urologist
Emotional problems with family and patient, Educational issues. Pressure areas, care of suprapubic catheter	Appropriate counselling. Information re support groups and respite care. Information about financial allowances is an important aspect of care. Management of health care at Home. Assistance and support of educational issues at home and in the school system, formal cognitive assessment.	Medical social workers, psychologists, special education teachers, nurses