Cerebral Palsy: Aetiology, Associated Problems and Management
Lecture for FRACP candidates
July 2010

What is cerebral palsy?

Definition (Bax 1964)
Cerebral palsy is a disorder of movement and posture due to a defect or lesion of the immature brain.

Cerebral palsy – an umbrella term
- All children are different
- The associated problems may be more significant than the motor disorder
- Permanent, non-progressive but not unchanging

New definition of cerebral palsy
Cerebral palsy describes a group of developmental disorders of movement and posture, causing activity restriction or disability, that are attributed to disturbances occurring in the fetal or infant brain. The motor impairment may be accompanied by a seizure disorder and by impairment of sensation, cognition, communication and/or behaviour (Rosenbaum 2007)
Storm’s words

Classification systems
- Nature of movement disorder
  eg spasticity, dystonia, ataxia
- Distribution eg diplegia, quadriplegia
- Severity

Why does classification matter?
- Descriptors for parents
- Type of treatment needed
- Prognosis
- Associated problems
- Information for service providers
- Research studies – drawing together similar groups

Type of motor disorder
- Spasticity
- Dyskinesia
- Athetosis
- Dystonia
- Ataxia
- Hypotonia
- Mixed

Distribution
- Quadriplegia
- Diplegia
- Hemiplegia
- Arms
- Legs
- Trunk
- Head and neck

Severity
- Mild
  - walks independently
- Moderate
  - walks with sticks / frame
- Severe
  - wheelchair dependent
**Classification of severity**

Gross Motor Function Classification System (The “GMFCS”)

**The GMFCS**

- Level 1 – walk without restrictions
- Level 2 – walk independently but more limitations
- Level 3 – need mobility devices eg frames
- Level 4 – sit on a regular chair, but use wheelchair
- Level 5 – no means of independent mobility

**Measuring motor progress**

The Growth Motor Curves
What causes cerebral palsy?

Case history of Lisa

- PMH
  - Pregnancy and birth normal
  - Crawled 10 months
  - Sat 12 months
- At 8 years
  - Presented to RCH
  - Falling more
  - Motor problems had deteriorated
- At 24 months
  - Diagnosis of spastic diplegia

History

Little: poor obstetric care associated with birth asphyxia responsible for most cases
Freud: adverse fetal events early in development may cause both birth complications and spasticity

Changing views

1960's: mechanism for the prevention of kernicterus
1970's: increased resources for obstetric and neonatal care, for example, increased Caesarean Section rate and use of electronic fetal monitoring

Results of changes in practice

No major change in cerebral palsy rates

Prevalence of cerebral palsy

2 per thousand live births
130 new cases in Victoria each year
Rates of CP, neonatal deaths and stillbirth rates Victoria, 1973 - 1999

Gender
- Males are over represented in all case series of cerebral palsy

Risk factors and causes: challenges

- Many risk factors, for example, prematurity, but few definite causes
  - Rates 25-30 x higher in infants weighing less than 1500 g
  - Babies who weigh <2500g account for 1/3 of children with cerebral palsy
- Causal pathways – a series of factors leading to the damaging event

Why do premature infants develop cerebral palsy?
1. Are they damaged before birth and then survive with good neonatal care?
2. Do they develop complications of prematurity such as IVH?

When does cerebral palsy occur?
- Antenatally 75%
- Perinatal 10 - 15%
- Postnatal 10%
- The cause remains unknown in a substantial proportion of cases, and is therefore attributed to antenatal events

Prenatal events
- Prenatal (75%)
  - Brain malformations eg cortical dysplasias
  - Intrauterine infections eg CMV
Prenatal events

- Vascular eg infarct in area of middle cerebral artery
- Metabolic eg iodine deficiency
- Toxic eg lead, mercury

Perinatal events

- Perinatal 10 - 15%
  - Hypoxic ischemic encephalopathy
  - Infection

"Birth asphyxia"

Even in the group where there appears to be evidence of asphyxia, it may not have been preventable

Post neonatal causes

- Postnatal 10%
  - Injury
    - accidental
    - non-accidental
  - Infections
    - meningitis
    - encephalitis

New directions

- The Australian Cerebral Palsy Register
  - A collaboration between all the States and the AIHW

- The Victorian Cerebral Palsy Register
  - Collects information about all children and young adults with cerebral palsy born in Victoria since 1970
**New directions**
- Role of thrombophilic mutations – Factor V Leiden mutation and other coagulopathies
- Maternal infection – urinary tract infection, chorioamnionitis
- Multiple pregnancy
- Low birth weight
- More information from MRI

**Prevention of cerebral palsy**
- This will only be achieved when more information is available about the multiple causes

**Diagnosis**
- Follow up of “at risk” infants, e.g., those of low birth weight
- Delayed motor development, eg, delay in learning to sit and stand
- Abnormalities of behaviour

**Early signs of cerebral palsy**
- Assymetric motor development
- Fisting of one hand
- Early favouring of one hand for reaching and grasping
- Assymetrical position in crawling
- “Limp” when starts to walk

**Early signs of cerebral palsy**
- Abnormalities of muscle tone
- Persistence of primitive reflexes
- Feeding problems
- Abnormalities of behaviour

**Difficulties in diagnosis**
- Different grades of severity
- Wide range of intellectual ability
- Signs may be late in appearance
- Prematurity
Collaborative perinatal project

- 229 children with abnormal signs during the first year of life
- At 7 years of age, 118 were free of motor handicap

Diagnostic approach

- MRI brain recommended (American Academy of Neurology Practice Parameter)
  - Genetic counseling
  - Establishing timing
- The majority of children will have abnormalities
  - 80-90% with MRI
  - 75% with CT

Diagnostic approach

Genetic and metabolic causes are unusual but should be considered in the presence of a normal MRI

- The true prevalence of these disorders in CP is not known
- Some may be treatable eg dopa responsive dystonia, glutaric aciduria, biotidinase deficiency
- Some may have associated problems that need treatment eg Leisch-Nyhan Disease

What can MRI tell us about timing?

- Brain malformations: 12 – 20 weeks
- Periventricular white matter injury: 26 – 34 weeks
- Cortical and subcortical gliosis: 36 – 44 weeks

Confirming the diagnosis

- A period of observation may be necessary
- Multidisciplinary assessment may be helpful

Issues in management

- Associated disabilities
- Health problems
- Consequences of the motor disorder
**Associated disabilities**

- **Epilepsy**
  - Occurs in about 40% of all children with cerebral palsy
  - Most common in spastic quadriplegia (50% - 94%) and hemiplegia (33% - 50%)
  - Higher incidence of refractory seizures and admissions for status epilepticus (Gururaj et al. Seizure 12:2;2003)
  - Few population based studies

- **Hearing problems**
  - Squints
  - Refractive errors
  - Field defects
  - Cortical visual impairment

- **Visual deficits**
  - Squints
  - Refractive errors
  - Field defects
  - Cortical visual impairment

- **Cognitive deficits**
  - Children may have intellectual, learning and perceptual problems. Assessment can be difficult in the presence of severe physical disability

- **Nutritional problems**
  - Under nutrition and failure to thrive
  - Difficulties with sucking and swallowing
  - Tongue thrusting
  - Decreased tongue movements and lip closure
  - Hypo or hypersensitive gag response
  - Meal times become long and carers anxious

- **Health problems - undernutrition**
  - 235 participants with GMFCS 111, 1V and V, average age 9.7 years
  - Indicators of malnutrition common
    - 47% of children had weight < 5% for age/gender
    - 68% had short stature
  - Correlated with increased health care utilisation and decreased participation in normal activities
  - (Samson-Fang et al J Pediatr 141;5:2002)

- **Gastrostomy**
  - Avoids aspiration
  - Often improves nutrition
  - Saves many hours of meal time assistance which can be devoted to other activities
Impact of quality of life on carers

At 12 months after gastrostomy placement, carers reported:

- Reduction in feeding times, increased ease of drug administration
- Reduced concern about their child's nutritional status
- Significant measurable improvement in quality of life of carers