Changing Perspectives in Cerebral Palsy

Dr Owen Hensey
Definition
Cerebral Palsy

A *persistent*, but not unchanging, disorder of movement and posture due to a *non-progressive* lesion in the *immature brain*
Proposed definition and classification of cerebral palsy: outcome of an international symposium

DMCN  April 2005: 47: 571-6
Proposed Definition of Cerebral Palsy

Cerebral palsy describes a group of disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain.

The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication, perception, and/or behaviour, and/or by a seizure disorder.
Cerebral Palsy

- Spastic: 80 - 90%
- Dyskinetic: 5 - 10%
- Ataxic: <5%
- Mixed
Aetiology
Aetiology of Cerebral Palsy

- Prenatal
- Perinatal
- Postneonatal
- Unknown
Brain development during gestation and early postnatal life
# Prenatal factors

**Periconceptional and early antenatal causes of the cerebral palsies**

<table>
<thead>
<tr>
<th>Type of factor</th>
<th>Description</th>
<th>Timing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maternal</td>
<td>Genetic, family history (<em>e.g.</em> epilepsy)</td>
<td>Periconceptional</td>
</tr>
<tr>
<td></td>
<td>Infertility, related problems</td>
<td>Periconceptional</td>
</tr>
<tr>
<td>Fetal malformation</td>
<td>Known and unknown genetic or teratogenic influences</td>
<td>Periconceptional and early gestation</td>
</tr>
<tr>
<td>syndromes</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Infections</td>
<td>TORCH, other organisms</td>
<td>Early gestation</td>
</tr>
<tr>
<td>Deficiencies</td>
<td>Iodine</td>
<td>Early-mid gestation</td>
</tr>
<tr>
<td></td>
<td>Thyroid hormone</td>
<td>?Throughout gestation</td>
</tr>
<tr>
<td>Toxic</td>
<td>Methylmercury</td>
<td>Throughout gestation</td>
</tr>
<tr>
<td></td>
<td>Alcohol</td>
<td>Early-mid gestation</td>
</tr>
<tr>
<td></td>
<td>Carbon monoxide poisoning</td>
<td>Throughout gestation</td>
</tr>
<tr>
<td>Vascular</td>
<td>Hypoxia, ischaemia, thrombotic disorders (<em>e.g.</em> maternal)</td>
<td>?Mid-late gestation</td>
</tr>
</tbody>
</table>
Picture no.1
Picture no.2
Schizencephaly
Schizencephaly

T2
Perinatal

- Hypoxia and birth asphyxia
- Infection
- Intracranial haemorrhage
- Metabolic
  - Hyperbilirubinaemia
  - Hypoglycaemia
Picture no.4
PVL

T2

T2 Flair
Spastic Diplegia
Fig. 7.3. Gestational-age-specific cerebral palsy rates by predominant cerebral palsy type, Western Australia 1980–92 combined (L. Watson and F. Stanley, unpublished data—excludes cases due to postneonatal causes).
Postneonatal

- Infection -
  - Cerebral
  - Gastroenteritis + dehydration
  - Pertussis
- Head injury -
  - Accidental
  - Non-accidental
  - Falls
- CVA -
  - Spontaneous
  - Postoperative
- Others -
  - ALTE
  - Near-drowning
  - Status epilepticus
Fig. 12.2. Laboratory reports of *Haemophilus influenzae* type b by age (England and Wales 1989–1998, provisional). (Data from Public Health Laboratory Service, Communicable Disease Centre.)
Epidemiology
Cerebral Palsies: Epidemiology and Causal Pathways

Stanley F, Blair E, Alberman E

Clinics in Developmental Medicine, No 151, 2000
Cerebral palsy and signs of birth asphyxia

Fig. 9.2. Possible pathways to cerebral palsy with signs of birth asphyxia.
Causal pathways: Preterm birth

* prolonged premature rupture of membranes

Fig. 7.5. Causes of preterm birth: which are on a causal pathway to cerebral palsy?
Causal pathways: IUGR

Fig. 8.5. The causes of intrauterine growth restriction.
Western Australia Cerebral Palsy Register

1980 - 1994
Prevalence of Cerebral Palsy

Fig. 4.3. Cerebral palsy rates per 1000 live births (excluding cases due to postneonatal causes) in four populations during the years 1959–1992 (except Mersey, 1967–1989).
Fig. 4.6. Low birthweight neonatal survivor rates in Western Australia, 1970–1995.
Fig. 4.7. Preterm cerebral palsy rates per 1000 neonatal survivors (excluding cases due to postneonatal causes) in Western Australia, 1980–1992.
Fig. 4.5. Very low birthweight (<1500 g) cerebral palsy rates per 1000 neonatal survivors (excluding cases due to postneonatal causes) in three populations (Sweden 1967–1990, Australia 1967–1992, Mersey 1967–1989).
Fig. 10.3. Rates of cerebral palsy (excluding cases due to postneonatal causes) in multiple and singleton births in Western Australia, 1960–1992, by five-year moving averages. (1992 rates are based on preliminary data for 1993–1994.)
Cerebral palsy following term newborn encephalopathy: a population-based study

Nadia Badawi* PhD MSc FRCP (I) FRACP, Department of Neonatology, The Children’s Hospital at Westmead, University of Sydney, Sydney, Australia;

Developmental Medicine & Child Neurology 2005, 47: 293–298
Western Australia 1993 - 1996

• 276 neonatal encephalopathy:
  25 died (9.1%)
  251 survivors -> 32 CP (13%)

• 99 term infants - CP
  - no encephalopathy
### Table II: Predominant type of term cerebral palsy according to history of encephalopathy in children born in Western Australia between June 1993 and December 1996

<table>
<thead>
<tr>
<th>Type of cerebral palsy</th>
<th>History of newborn encephalopathy (n=32)</th>
<th>No history of newborn encephalopathy (n=99)</th>
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<tbody>
<tr>
<td>Spastic quadriplegia</td>
<td>10 (31)</td>
<td>12 (12)</td>
</tr>
<tr>
<td>Spastic diplegia</td>
<td>6 (19)</td>
<td>24 (24)</td>
</tr>
<tr>
<td>Hemiplegia</td>
<td>7 (22)</td>
<td>38 (38)</td>
</tr>
<tr>
<td>Athetosis/dystonia</td>
<td>8 (25)</td>
<td>14 (14)</td>
</tr>
<tr>
<td>Ataxia/hypotonia</td>
<td>1 (3)</td>
<td>11 (11)</td>
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Numbers in parentheses are percentages.
Western Australia 1993 - 1996

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Numbers in parentheses are percentages.
Western Australia 1993 - 1996

Figure 1: Severity of cerebral palsy (CP) in children born at term in Western Australia between June 1993 and December 1996 (■) with and (□) without newborn encephalopathy.
Western Australia 1993 - 1996

Figure 2: Type of additional disability in children with cerebral palsy (CP) born at term in Western Australia between June 1993 and December 1996 with (■) and without (□) newborn encephalopathy.
Surveillance of Cerebral Palsy in Europe (SCPE)
16 SCPE centres
SCPE: Birthweight Distribution of Cerebral Palsy Cases 1980 - 1996

- <1500g: 52.1%
- 1500 - 2499g: 21.1%
- >2500g: 26.8%

Prevalence of CP: 2.03/1000 livebirths
Surveillance of Cerebral Palsy in Europe (SCPE)

Trends in cerebral palsy among infants of very low birthweight (<1500 g) or born prematurely (<32 weeks) in 16 European centres: a database study

Mary Jane Platt, Christine Cans, Ann Johnson, Geraldine Surman, Monica Topp, Maria Giulia Torrioli, Inge Krageloh-Mann

Lancet 2007; 368 : 43 - 50
SCPE 1980 - 1996

- 1575 VLBW infants born with CP
  - <1000g: 414 (26%)
  - 1000g – 1499g: 1161 (74%)

- Spastic CP: 94%

- VLBW birth prevalence:
  - 1980: 60.6/1000 livebirths
  - 1996: 39.5/1000 livebirths
SCPE : 1980 – 1996
Cerebral Palsy Rates

<1000gms

1000 – 1499gms

*Lancet 2007; 368 : 43 - 50*
SCPE : 1980 – 1996
CP Subtypes

Figure 4: Birthweight-specific birth prevalence of bilateral and unilateral spastic cerebral palsy (3-year moving average) from nine European centres, 1980-96. Error bars= SE.

Lancet 2007; 368 : 43 - 50
SCPE: 1980–1996

Gestational Age

Figure 5: Gestational-age-specific birth prevalence of cerebral palsy (3-year moving average), from nine European centres, 1980–96. Error bars=SE.

Lancet 2007; 368: 43-50
SCPE 1980 - 1996

Conclusion:

Survival of infants of birthweight <1500gms continues to improve and this improvement is not associated with increased morbidity.

- Overall prevalence: 2.2/1000 livebirths

- <1500 gms:
  - 1984 – 6: 64.2/1000 livebirths
  - 1987 – 90: 90.5/1000 livebirths
  - 1991 - 1997: 44.5/1000 livebirths

- Decreased severity of motor and intellectual impairment
Figure 1: Numbers of children with cerebral palsy by individual birth year and birthweight group, 1981 to 97 (excluding 1988).

Prevalence 2.2/1000 livebirths
Medical Management
Reproduced from Graham & Selber, 2003
Mechanism of spasticity

Fig. 1. Muscle tone is regulated by output from α motoneurons, the output of which is influenced by: (i) excitatory afferent impulses from skeletal muscle; and (ii) inhibitory impulses descending from the brain. The latter impulses are deficient in patients with spasticity of cerebral origin. Abbreviation and symbols: GABA = γ-aminobutyric acid; α = α motoneuron; + indicates an excitatory impulse; – indicates an inhibitory impulse.
Treatment approaches to spasticity

- Physical methods
- Pharmacological agents
- Surgery - orthopaedic
- Neurosurgical
Physical methods

Bobath therapy
Casting
Orthotics
Electrical stimulation
Pharmacological

**Oral**
- Diazepam
- Baclofen
- Dantrolene
- Tizanidine

**Local**
- Phenol
- Botulinum toxin A

**Intrathecal**
- Baclofen
Oral medication

Pathways involved in spasticity

Baclofen
Diazepam
Tizanidine

Dantrolene
BTX-A
Local

Phenol

Botulinum toxin A
Botulinum Toxin A: Mode of Action

1. BTX-A
2. Exocytotic vesicle containing acetylcholine
3. Blocked exocytotic vesicle
Injection Sites

- Gastrocnemius
- Proximal, superficial site
- Distal, deeper site
- Tibia
- Soleus
Safety profile

Adverse events (7%)

Local: weakness (1%)
pain

Generalised: weakness / falls
urinary / faecal incontinence (1%)
skin rash
aspiration

Systemic: fatigue
flu-like symptoms

Bakheit et al, DMCN 2001, 43: 234-238
Orthopaedic Surgery

- Mature gait
- Conservative methods no longer effective
- Before bony deformity develops
Neurosurgical approaches

Intrathecal baclofen

Selective dorsal rhizotomy
Intrathecal baclofen
Intrathecal baclofen

Complications:

- Infection
- Catheter leaks / breaks
- Overdose

- Hypotonia (25%)
- Drowsiness (20%)
- Dizziness (8%)
- Paraesthesia (6.7%)
- Convulsions (4.7%)
- Urinary retention (1.9%)
Benefits of ITB

• Maintains long-term reduction in spasticity
• May reduce painful muscle spasms
• Reduces discomfort due to difficulties with positioning
• Reversible
Selective dorsal rhizotomy
Selective dorsal rhizotomy

**Vancouver:** Steinbok P, et al  
DMCN 1997, 39:178-84

**Seattle:** McLaughlin JF, et al  
DMCN 1998, 40:220-32

**Toronto:** Wright FV, et al  
DMCN 1998, 40:239-47
# Controlled trials

<table>
<thead>
<tr>
<th></th>
<th>No. patients</th>
<th>Duration of Physiotherapy</th>
<th>Follow-up</th>
</tr>
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<tbody>
<tr>
<td>Vancouver:</td>
<td>28</td>
<td>9m</td>
<td>9m</td>
</tr>
<tr>
<td>Seattle:</td>
<td>38</td>
<td>12m</td>
<td>24m</td>
</tr>
<tr>
<td>Toronto:</td>
<td>24</td>
<td>12m</td>
<td>12m</td>
</tr>
</tbody>
</table>
Controlled trials

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Studies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reduction in spasticity</td>
<td>All</td>
</tr>
<tr>
<td>Improvement in motor function in rhizotomy group</td>
<td>Vancouver</td>
</tr>
<tr>
<td>Rhizotomy does not lead to greater functional gains than intensive physiotherapy</td>
<td>Toronto, Seattle</td>
</tr>
</tbody>
</table>
Management of spasticity

- Oral therapy
- ITB
- SDR
- BTX-A
- Surgery

Focal

Reversible

General

Permanent

BTX-A = Botox®
ITB = intrathecal baclofen
SDR = selective dorsal rhizotomy
Will he walk?

Picture no.5

- Premature-born at 29 weeks.
- Ventilated for 3 days.
- Ultra sound in early days was normal.
- Referred to C.R.C at 17 months.
- Physiotherapy began twice weekly and home programme.
2 years and 6 months

Picture no.6

- Prepositioning of limb
- Stability in stance
- Foot clearance in swing
- Adequate step length
- Energy conservation
  (Jim Gage)
Age 3+ years

Picture no.7

- Prepositioning of limb
- Stability in stance
- Foot clearance in swing
- Adequate step length
- Energy conservation
  (Jim Gage)
5 years of age

Picture no.8

7 years of age

Picture no.9
Concerns

- **Parents** - “how he looks among his peers and to do the right thing for him”

- **Physiotherapist** - “his gait pattern, the effect independent walking has on his joints, muscle weakness”

- **Doctor** - “internal rotation gait”
Surgery.

• Bilateral femoral derotation osteotomies with distal medial hamstring lengthening.

• Intensive rehab programme post op, 50 hours in first 6 months.
Pre Op

Picture no.10

Post Op

Picture no.11
How can we preserve the gains achieved while attending to his present need for independent walking?

Picture no. 12
The good news is you’re about to be attracted to girls!

The bad news is you’ll be covered in pimples.
Effects of puberty

Disproportionate bone-to-muscle growth
Increased weight gain
Asymmetry
Weakness
Non-compliance
Conservative/Non Operative Treatment Modalities

Surgical Management

Primary

Secondary & Tertiary

CP

0  2-7 yr  16 yr  30 – 40 yr  50 yr  75 yr

Walk

Growth stops

Physiotherapy
Orthotics
Casting
Botulinum toxin
Walking Aids
Medical

Walking Aids
Survival
Cerebral palsy: why we must plan for survival

<table>
<thead>
<tr>
<th>Year</th>
<th>Authors</th>
<th>Location</th>
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<tbody>
<tr>
<td>1990</td>
<td>Evans PM et al</td>
<td>SE Thames, UK</td>
</tr>
<tr>
<td>1994</td>
<td>Hutton JL et al</td>
<td>Mersey Region, UK</td>
</tr>
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<td>1995</td>
<td>Crighton JU et al</td>
<td>Br. Columbia, Canada</td>
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<tr>
<td>1998</td>
<td>Strauss D, Shavelle RM</td>
<td>California, USA</td>
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<td>2000</td>
<td>Hutton JL et al</td>
<td>NE England</td>
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<tr>
<td>2001</td>
<td>Blair E et al</td>
<td>W Australia</td>
</tr>
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</table>
20 year survival

Evans et al, 1990
Survival for differing types of cerebral palsy

Evans et al, 1990
Life expectancy in cerebral palsy

Ambulation
Manual dexterity
Mental ability
Feeding ability
Percentage survival of subjects with cerebral palsy

<table>
<thead>
<tr>
<th>Age (yrs)</th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>3</th>
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<tbody>
<tr>
<td>10</td>
<td>99.7</td>
<td>96.5</td>
<td>92.1</td>
<td>70.0</td>
</tr>
<tr>
<td>15</td>
<td>99.0</td>
<td>95.5</td>
<td>90.4</td>
<td>54.5</td>
</tr>
<tr>
<td>20</td>
<td>98.8</td>
<td>93.5</td>
<td>84.7</td>
<td>50.3</td>
</tr>
<tr>
<td>25</td>
<td>98.8</td>
<td>90.6</td>
<td>80.5</td>
<td>44.2</td>
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</table>

*Hutton et al, 1994*
Survival after 15 years

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Mortality Risk</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tube fed</td>
<td>8.2</td>
</tr>
<tr>
<td>Cannot lift head</td>
<td>5.9</td>
</tr>
<tr>
<td>Does not walk</td>
<td>4.0</td>
</tr>
<tr>
<td>Severe cerebral palsy</td>
<td>2.9</td>
</tr>
<tr>
<td>Profound learning disability</td>
<td>2.9</td>
</tr>
<tr>
<td>Non-verbal</td>
<td>2.8</td>
</tr>
</tbody>
</table>

Strauss, Shavelle, 1998
Life expectancy among people with cerebral palsy in Western Australia

Blair et al
DMCN 2001, 43: 508-515
Life expectancy among people with cerebral palsy in Western Australia

Figure 3: Survival by severity of CP.
Levels of severity:
- Minimal
- Mild
- Moderate
- Severe

Blair et al, DMCN 2001, 43: 508-515
Life expectancy among people with cerebral palsy in Western Australia

Figure 2: Survival by intellectual ability.

Blair et al, DMCN 2001, 43: 508-515
Improved survival

Respiratory care
Gastrostomy feeding
Epilepsy control

Improved nutrition
Immunization
Improved social and living standards
Survival in Cerebral Palsy in the last 20 years: signs of improvement?

California : 1983 – 2002

- Mortality rates in those children with the most severe disabilities, and of adults who are fed by gastrostomy fell by 20% over the 20-year period

- In these groups life expectancies rise by approximately 5 years
