Cerebral palsy in childhood

Little’s Disease
Definition of Cerebral Palsy (2006)

“Cerebral palsy describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing foetal or infant brain. The motor disorders of CP are often accompanied by disturbances of sensation, perception, cognition, communication and behavior, by epilepsy, and by secondary musculoskeletal problems”.
Developmental perspective

- Although the lesion itself is non-progressive, the clinical picture will be modified by age.
- Evolving medical and psychosocial issues at each stage of life
- Incidence in well resourced countries relatively unchanged
- Incidence in SA unknown
Clinical Presentation

- Delayed motor milestones
- Asymmetry of posture or movement
- Persistent primitive reflexes
- Delayed development of protective reflexes
Asymmetry of posture/movement
ATNR: Asymmetric tonic neck reflex
Lateral protective reflex
Spastic quadriplegia with fisting and scissoring
# UMN syndrome

**Positive signs:**
- increased tone in spastic CP
- overactive tendon reflexes
- clonus

**Negative signs:**
- weakness
- selective muscle control
- easy fatigability
- poor dexterity
- poor balance
These patients have Cerebral Palsy
APPROACH

1. Is it CP?
2. What type?
3. What is the cause?
4. How severe is motor impairment?
5. What is the child’s cognitive potential?
6. What are the associated problems?
7. Short and long term management goals?
Is it CP?

It may not be CP if there is:

1. Regression
2. Diurnal variation in function
3. Dystonia
4. Predominant weakness

Exclude the mimics!
What type of CP?

Classification

- Motor: abnormalities of tone or movement
- Topographical: Distribution of the defect
- NB not always that simple
Motor Abnormality

- Spastic → Increased tone (77-93%)
- Dyskinetic → Abnormal movements (2-15%)
- Ataxic → Unsteadiness (2-8%)
- Hypotonic → Uncommon (0.7-2.6%)
- Mixed → Spasticity with movement disorder.
Topographical Classification by Distribution

- Diplegia
- Hemiplegia
- Quadriplegia

Less Affected Areas
More Affected Areas
Classification by Distribution
3. What is the cause?
Aetiological classification

Insult to the motor cortex or motor pathways of the immature brain (before 2-5 years)

Prenatal

Perinatal

Postnatal

Often multiple risk factors
- **Prenatal risk factors**
  - Intra uterine infection e.g. TORCH, chorioamnionitis
  - Intra uterine growth retardation
  - Vascular incidents  Brain malformations Non-cerebral anomalies
  - Toxic agents e.g. methyl mercury, alcohol, drugs
  - Genetic : inherited thrombophilias, parental consanguinity
  - Maternal factors: diabetes, hypothyroidism, pre-eclampsia, antepartum hemorrhage, thrombophilia, recurrent miscarriages
  - Multiple pregnancies (monochorionic)
  - Co-fetal death
  - Poor socioeconomic status

- **Perinatal factors**
  - Birth asphyxia, birth injury
  - Prematurity, neonatal septicemia and meningitis
  - Hypoglycaemia, hyperbilirubinemia, Transient neonatal hypothyroxinaemia

- **Postnatal factors**
  - Meningitis esp TBM, Trauma, vascular episodes.
Correlation between specific insult and type of Cerebral palsy exists in some cases:

- Severe HIE in term infant $\rightarrow$ Spastic quadriplegia
- Premature infant with IVH/PVL $\rightarrow$ Spastic diplegia
- Neonatal hypoglycaemia $\rightarrow$ Ataxic cerebral palsy
- Severe kernicterus $\rightarrow$ Choreo-athetoid cerebral palsy
- TB Meningitis with infarct $\rightarrow$ hemiplegia
What is the cause?

- MRI preferred modality (affected areas, timing of insult, and cause)
  1. Early brain malformations (10%)
  2. White matter injury
  3. Neonatal encephalopathies
  4. Postnatally acquired disorders (heterogeneous group)
Neuroimaging

- Pathology unidentified in 10-20% (often ataxic CP)
- Accurate prediction of CP difficult
- In most cases aetiological diagnosis does not change recommendations for child’s care.
- Exceptions are inborn errors of metabolism (2%)
Glutaric aciduria Type 1
How severe is it?

- Gross Motor Function Classification System (GMFCS-ER) Includes 5 levels of ambulatory ability in children up to 18 years
  Describes each level across four age bands

- Manual Ability Classification system (MACS)

- Communication Function Classification system (CFCS)
  NB Not sensitive enough for evaluating interventions
GMFCS E & R between 6th and 12th birthday: Descriptors and illustrations

**GMFCS Level I**
Children walk at home, school, outdoors and in the community. They can climb stairs without the use of a railing. Children perform gross motor skills such as running and jumping, but speed, balance and coordination are limited.

**GMFCS Level II**
Children walk in most settings and climb stairs holding onto a railing. They may experience difficulty walking long distances and balancing on uneven terrain, inclines, in crowded areas or confined spaces. Children may walk with physical assistance, a hand-held mobility device or used wheeled mobility over long distances. Children have only minimal ability to perform gross motor skills such as running and jumping.

**GMFCS Level III**
Children walk using a hand-held mobility device in most indoor settings. They may climb stairs holding onto a railing with supervision or assistance. Children use wheeled mobility when traveling long distances and may self-propel for shorter distances.

**GMFCS Level IV**
Children use methods of mobility that require physical assistance or powered mobility in most settings. They may walk for short distances at home with physical assistance or use powered mobility or a body support walker when positioned. At school, outdoors and in the community children are transported in a manual wheelchair or use powered mobility.

**GMFCS Level V**
Children are transported in a manual wheelchair in all settings. Children are limited in their ability to maintain antigravity head and trunk postures and control leg and arm movements.

Fig. 3. Age-specific example of criteria for the Gross Motor Function Classification System (GMFCS). (Courtesy of Kerr Graham, MD, The Royal Children's Hospital, Melbourne, Australia.)
What is child’s cognitive potential?

- Spastic quadriplegia → high association with intellectual disability.
- Spastic hemiplegia → 25% cognitive abnormalities.
- Choreo-athetoid cerebral palsy → Intellect preserved in many cases.
- Spastic diplegia → good prognosis for normal intellectual development.
Cerebral palsy student defies own expectations with eight distinctions

NALISHA KALIDEEN

JOHANNESBURG: Benjamin Rosman, 18, was born with cerebral palsy and specialists didn't believe he'd cope with normal schooling but he has proved them wrong, getting eight distinctions in his matric exams.

The Crawford College, Sandton pupil even defied his own expectations by getting a distinction in history, a subject he wasn't sure he passed because when he wrote the exam he was suffering from a heavy bout of flu.

The subjects he received distinctions for are English, Afrikaans, maths, physical science, computer studies, additional maths, history and art.

Rosman is partially paralysed on his right side. He has limited control of his right hand and walks with a limp.

"I've always tried not to think of it as a disability. There are people with whom I've been to school with for years who didn't even know I had cerebral palsy," Rosman said.

His mother, Minessa, thinks that the key to her son's success has been the fact that he's never thought of himself as being either different or handicapped in any way.

"We never treated him as if he was handicapped. We've always told..."
What are the Associated problems?

1. Sensory
2. Cognitive
3. Physiological

Number of associated impairments increase with severity of motor impairment
Sensory impairments

- Visual problems (21-63%) myopia, amblyopia, visual field deficits, cortical blindness, retinopathy of prematurity, strabismus
- Hearing impairment (10%-25%)
- Somatosensory deficits
Physiological

- Epilepsy (40%)
- Failure to thrive
- Feeding problems
- Gastro-esophageal reflux
- Constipation
- Dental
- Spasticity
- Orthopaedic
- Behavioural
Principles of Management
Primum non nocere

- Interventions must lead to improved functional outcomes
- Prevention of secondary problems e.g. hip dislocation, dental caries, constipation, rickets
- Prioritise communication, activities of daily living, mobility, participation
Prevention of Hips deformities

- Outcome depends on ambulation
- Incidence of hip displacement
  - GMFCS level 1 0%
  - GMFCS level V 90%
- Systematic surveillance in spastic CP from 12-18 months of age
- 6 monthly hip Xrays if GMFCS 111-V
Management of spasticity

- Aim is to maintain musculoskeletal health into adulthood
- Oral medications: diazepam, baclofen
- Intramuscular medications: botox, phenol
- Neurosurgical: selective dorsal rhizotomy, intrathecal baclofen
Management of dystonia

- Generalised dystonia: trial of carbidopa/levodopa 10/100mg bd, increasing to 25/100mg tds
- Trial of anticholinergics; trihexiphenidyl (benzhexol) 2.5mg/day, increase to max 15mg daily
- Trial of baclofen 10mg nocte/ IT pump
- Trial of benzodiazepines e.g. diazepam
- Focal dystonia: botox
Principles of Management

- Team approach
- Multidisciplinary
- Interdisciplinary
- Transdisciplinary
- Family-centred
Disciplines

- Primary Physicians: GPs/Paediatricians
- Developmentalists /Neurologists
- Orthopaedic surgeons NB involve early
- Social work
- Physiotherapy
- Occupational therapy
- Speech therapist
- Orthotist
- Dietician
- Nursing
Other Disciplines

- Neurosurgery
- Gastroenterology
- ENT
- Dentistry
- Ophthalmologist
- Education
- Augmentative communication
- Psychology
- Vocational counsellor
ADVANCES

- Importance of Nutrition
- CP growth charts for spastic CP
- Vitamin D supplements
- Referral for gastrostomy feeding tubes when indicated
Assistive devices

• High tech / Low tech

N.B. Practicality and affordability

• Low technology usually includes devices that are passive or simple, with few moving parts
  • Picture boards
  • Magnifying screens
  • Built-up handle spoons
  • Reachers
  • Side-positioners
- Designed to provide postural support while lying down.
- Inhibits abnormal reflex patterns, extreme flexion and extension.
- Used in conjunction with seating and standing devices to provide 24 hour postural support.
- Removable washable covers and waterproof base cover.
- Alternative customised back positioner available when side lying is not recommended.

Visit our website for more detailed information

- One unit fully adjustable for prone, upright and supine standing.
- Adjustable gas strut allows tilt between 90 and 180 degrees.
- Height adjustable full body supports and laptray.
- Mobile with braking caster wheels.
- Recommended for improving pelvic and trunk control.
- Encourages weight bearing and reduces tone in the lower limbs.
- Provides stretch to Achilles tendons and hamstrings.
SWASH (Sitting Walking And Standing Orthosis)
SWASH

- Hip stabilisation in sitting and walking
- Less hip abduction
- More upright posture
- Benefits both ambulatory and non-ambulatory child
- Mild to severe CP
Assistive devices

• High technology includes devices that have greater complexity and may have an electronic component
  • Computers
  • Motorised wheelchairs
  • Environmental control units
  • Speech recognition software
Gait deviations in CP

- Primary
  - spasticity
  - motor control
  - balance
  - weakness
- Secondary musculoskeletal problems
  - contractures
  - bone deformities (eg femoral anteversion)
  - joint instability
- Tertiary compensations
Role of Gait Analysis

- Understanding biomechanics
- Diagnosis and treatment planning
- Assessing outcome
Advances

- **Orthopaedic surgery**: Single event multilevel surgery has replaced the “Birthday syndrome”

- **Neurosurgical Procedures**: Baclofen pumps for spasticity, Selective dorsal rhizotomy
Family-centred care: a more equal partnership

Families determine interventions that are appropriate based on knowledge of their child and their own circumstances and they determine timing of interventions.
Future Research

- No proven treatments to enhance brain function

- Three areas of current interest
  - Replace non-functioning brain cells
  - Repair cell connectors
dendrites and axons
  - Promote alternative brain pathways
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