Brain Injury Group

Specialist training from brain injury experts

Cerebral Palsy

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Welcome from the Chair

Andrew Hannam
Enable Law
Introduction to Cerebral Palsy

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Cerebral palsy

Charlie Fairhurst
Evelina London Children’s Hospital
Health Condition
(Disorder or Disease)

Body Functions and structures

Activities

Participation

Health Condition
(Disorder or Disease)

Body Functions and structures

Activities

Participation

Environmental factors

Personal factors

International Classification of Functioning, Disability and Health
2011
Cerebral palsy (CP) 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.”
NICE SPASTICITY CG145

- Spasticity in under 19s: management
- NICE guideline
  Published: July 2012
- nice.org.uk/guidance/cg145
Spasticity in children and young people overview

- Child or young person with spasticity
  - Principles of care
  - Service organisation
  - Treatment
    - Monitoring
      - Physical therapy
      - Orthoses
      - Pharmacological therapy
      - Surgery
        - Oral drugs
        - Botulinum toxin type A
        - Continuous pump-administered intrathecal baclofen
Cerebral palsy in under 25s: assessment and management

NICE guideline
Published: 25 January 2017

nice.org.uk/guidance/ng62
MOTOR FUNCTION

• CAREFUL BALANCE OF:
  – Neurological control
  – Posture
  – Reflex excitability
    • Nerves
    • Muscle
  – Biomechanics
    • Growth changes
Cerebral palsy

• Description not diagnosis
• Site of lesion > clinical signs
• **Classification** – types of motor impairment
  – Pattern of limb involvement - topography
  – Pattern of abnormal movement or muscle tone
  – Functional ability

• **Classification**
  – Site of lesion
  – Patterns of damage
Cerebral Palsy

- **Type** – spastic, dyskinetic or ataxic CP
- **Distribution** – bilateral or unilateral
- **Severity** – GMFCS Level I-V
- **Comorbidity** –
  - epilepsy,
  - other medical
  - global delay,
  - Behaviour,
  - sensory impairment
Upper Motor Neurone Syndrome

+ 
- 

- **Overactivation**
- **Spasticity**
  - increased muscle tone
  - hyperreflexia
  - clonus
  - abnormal co-contraction
- **Released flexor reflexes**
  - mass synergy patterns

- **Reduction in motor activity**
- **Loss of dexterity**
- **Weakness**
  - inadequate force generation
  - slow movements
- **Loss of selective control**

More amenable to intervention

Usually more disabling but less focussed on
Dystonia

Lesion in locomotor driving system

- Pattern of sustained disturbed muscle contractions causing abnormal postures.

- Frequently associated with involuntary movements
Cerebral Palsy

- **Type** – spastic, dyskinetic or ataxic CP
- **Distribution** – bilateral or unilateral
- **Severity** – GMFCS Level I-V
- **Comorbidity** –
  - epilepsy,
  - other medical
  - global delay,
  - Behaviour,
  - sensory impairment
GMFCS

GMFCS Level I

GMFCS Level II

GMFCS Level III

GMFCS Level IV

GMFCS Level V
I. Handles objects easily and successfully.

II. Handles most objects but with somewhat reduced quality and/or speed of achievement.

III. Handles objects with difficulty; needs help to prepare and/or modify activities.

IV. Handles a limited selection of easily managed objects in adapted situations.

V. Does not handle objects and has severely limited ability to perform even simple actions.
This graph shows the observed and predicted GMFM-66 scores for children in GMFCS Levels I through V. The curved solid lines indicate average performance. The horizontal dotted lines on the right of the figure indicate the band expected to encompass 50% of children's limits of development. The solid vertical lines indicate the average age-90 (the age in years by which children are expected to reach 90% of their motor development potential). The dotted vertical lines indicate the bands expected to encompass 50% of age-90 values around the average. The absence of 50% bands in level IV and level V indicates low variation in age-90 values.

Cerebral Palsy - aetiology

• Brain development

• Lesions of developing brain
  • antenatal, perinatal and postnatal

• Causes of lesions

• Classification

• Loopholes
BRAIN GROWTH

• Birth  average 400g
• Increased size
  • Continued synaptic connections
  • neuroglia, myelin
• Maximum rate of growth
  • In utero - 20 weeks postnatal
• Age 3  1200g
• Plateau age 18  m 1100 - 1700 (1360)
  f  1050 - 1550 (1275)
Epidemiology of Cerebral Palsy

- Most prevalent cause of motor disorders in childhood
  - Massive socio-economic impact
- Prevalence 2-3 per 1000 live births
- Increases up to 100 per 1000 live births in cases of extreme prematurity < 28/40

- 16 European centres – SCPE – 1575 infants
- Birth prevalence fell from 60.6 per 1000 (1981) very low birth weight (VLBW <1000g) infants to 39.5 per 1000 (1996)
Person under 25 with suspected cerebral palsy

- Identification and diagnosis
- Information and support
- Discussing prognosis
- Management
- Transition from children to adult services

- Organisation of care
- Patient and service user experience
Identifying and diagnosing cerebral palsy in under 25s

Person under 25 with suspected cerebral palsy

Risk factors

When to provide an enhanced follow-up programme

Signs of cerebral palsy

Red flags for other neurological disorders

When to refer to a child development service

Assessing causes

Multidisciplinary assessment and access to multidisciplinary care

Information and support
Risk factors

Recognise the following as independent risk factors for cerebral palsy:

• antenatal factors:
  • preterm birth, with risk increasing with decreasing gestational age
  • chorioamnionitis
  • maternal respiratory tract or genito-urinary infection treated in hospital

• perinatal factors:
  • low birth weight
  • chorioamnionitis
  • neonatal encephalopathy
  • neonatal sepsis (particularly with a birth weight below 1.5 kg)
  • maternal respiratory tract or genito-urinary infection treated in hospital

• postnatal factors:
  • meningitis.
Signs of cerebral palsy

Recognise the following as possible early motor features in the presentation of cerebral palsy:

• unusual fidgety movements or other abnormalities of movement, including asymmetry or paucity of movement
• abnormalities of tone, including hypotonia (floppiness), spasticity (stiffness) or dyskinesia (fluctuating tone)
• abnormal motor development, including late head control, rolling, and crawling
• feeding difficulties.

Recognise that the most common delayed motor milestones in children with cerebral palsy are:

• not sitting by 8 months (corrected for gestational age)
• not walking by 18 months (corrected for gestational age)
• early asymmetry of hand function (hand preference) before 1 year (corrected for gestational age).

If there are concerns that a child may have cerebral palsy but a definitive diagnosis cannot be made, discuss this with their parents or carers and explain that an enhanced clinical and developmental follow-up programme will be necessary to try to reach a definite conclusion.
CAUSE

Use of MRI
Assessing Cause
White matter damage
Congenital malformation
HIE
Post natal
Cumulative
IMAGING - MRI
Using MRI to assess cause

Offer MRI to investigate aetiology in a child or young person with suspected or known cerebral palsy if this is not clear from:

- antenatal, perinatal and postnatal history
- their developmental progress
- findings on clinical examination
- results of cranial ultrasound examinations.

Recognise that MRI will not accurately establish the timing of a hypoxic-ischaemic brain injury in a child with cerebral palsy.

When deciding the best age to perform an MRI scan for a child with cerebral palsy, take account of the following:

- Subtle neuro-anatomical changes that could explain the aetiology of cerebral palsy may not be apparent until 2 years of age.
- The presence of any red flags for a progressive neurological disorder (see red flags for other neurological disorders).
- That general anaesthesia or sedation is usually needed for young children having MRI.
- The views of the child or young person and their parents or carers.
Assessing causes

When assessing the likely cause of cerebral palsy in a [child], recognise that a number of MRI-identified brain abnormalities have been reported at the following approximate prevalences in children with cerebral palsy:

- white matter damage: 45%
- basal ganglia or deep grey matter damage: 13%
- congenital malformation: 10%
- focal infarcts: 7%.

When assessing the likely cause of cerebral palsy, recognise that white matter damage, including periventricular leukomalacia shown on neuroimaging:

- is more common in children born preterm than in those born at term
- may occur in children with any functional level or motor subtype, but is more common in spastic than in dyskinetic cerebral palsy.

When assessing the likely cause of cerebral palsy, recognise that basal ganglia or deep grey matter damage is mostly associated with dyskinetic cerebral palsy.

When assessing the likely cause of cerebral palsy, recognise that congenital malformations as a cause of cerebral palsy:

- are more common in children born at term than in those born preterm
- may occur in children with any functional level or motor subtype
- are associated with higher levels of functional impairment than other causes.
Recognise that the clinical syndrome of neonatal encephalopathy can result from various pathological events, such as a hypoxic-ischaemic brain injury or sepsis, and if there has been more than 1 such event occurs they may interact to damage the developing brain.

When assessing the likely cause of cerebral palsy, recognise that neonatal encephalopathy has been reported at the following approximate prevalences in children with cerebral palsy born after 35 weeks:

- attributed to a perinatal hypoxic-ischaemic injury: 20%
- not attributed to a perinatal hypoxic-ischaemic injury: 12%.

Recognise that for cerebral palsy associated with a perinatal hypoxic-ischaemic injury:

- the extent of long-term functional impairment is often related to the severity of the initial encephalopathy
- the dyskinetic motor subtype is more common than other subtypes.

Recognise that for cerebral palsy acquired after the neonatal period, the following causes and approximate prevalences have been reported:

- meningitis: 20%
- other infections: 30%
- head injury: 12%.

When assessing the likely cause of cerebral palsy, recognise that independent risk factors:

- can have a cumulative impact, adversely affecting the developing brain and resulting in cerebral palsy
- may have an impact at any stage of development, including the antenatal, perinatal and postnatal periods.
Aetiology of Cerebral Palsy

When it happens > what pattern seen

• **1st and 2nd trimester**
  • brain mal-developments

• **Early 3rd trimester - prematurity**
  • Intraventricular haemorrhage (IVH)
  • Periventricular leucomalacia (PVL)

• **Late 3rd trimester onwards**
  • Hypoxic-ischaemic events
  • Cortical - subcortical and deep grey matter lesions
Clinical Motor features with time

- Loss of mobility
- Lack of postural control
- Loss of dexterity
- Loss of bulbar control

- PAIN

- Care and nursing difficulties – Activities of daily living
ACTIVE ROLE
OPERATIONAL LEVEL
TAKING PART
EXPERIENCE
DEPENDENCE
SECURITY
ATTACHMENTS
COMFORT
PAIN FREE
SELF IDENTITY
ADJUSTMENT
CONTROL OF SELF
CHOICES
INDEPENDENCE

1. Uncontrolled
2. Experiencing
3. Taking Part
4. Active Role
5. Independence
Cerebral Palsy - review

• Normal neurology → Abnormal neurology
• Site of lesion leads to specific clinical picture
  – Different descriptions
    • USA
    • USE
• Range of Disability dependent on severity
  – Mobility, Posture, Activities of Daily Living
  – Pain, Spasm, Biomechanical change
  – Communication
  – Co-morbidities
  – Function, function, function

Relevance to patient ??
Summary

Each child is a dynamic individual

- Their needs and abilities should be regularly re-appraised
- They should not be stressed to perform inappropriate tasks
- Their physical and psychological core needs of comfort and security should be paramount to all members of their multidisciplinary team
Breach of duty - labour

Professor Tim Draycott
Southmead Hospital
SEE SEPARATE DOWNLOAD
Cerebral Palsy
Breach of Duty / Causation issues from a medical perspective

Simon Mitchell
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Brain Injury Group cerebral palsy training
London 12th September 2018
Cerebral Palsy

- Non-progressive disorder of movement & posture due to abnormal development or injury to motor control centres in the brain
- Mainly originating in prenatal or perinatal period
- Commonly associated disability
  - Cognitive impairment
  - Sensory impairments (vision, hearing)
  - Seizure disorders
- Many factors in aetiology
Origins of Cerebral Palsy

- **Intrauterine**
  - Primary cerebral malformation
  - Congenital Cytomegalovirus
  - Infection / inflammation
  - Hypoxic ischaemic injury
  - Chorioamnionitis
  - Placental insufficiency
  - Intrapartum hypoxic ischaemia
  - Fetomaternal haemorrhage

- **Stroke**
  - Arterial ischaemic stroke
  - Cerebral venous sinus thrombosis
  - Haemorrhage / trauma

- **Infection**
  - Neonatal meningitis
  - Viral meningitis / encephalitis

- **Others**
  - Hyperbilirubinaemia
  - Hypoglycaemia
Preterm infants (1)

• **Intraventricular / periventricular haemorrhage**
  • Germinal matrix haemorrhage
  • Highly vascular area prior to around 34 weeks gestation
  • Fragility of blood vessels confers vulnerability to bleeding
  • IVH or periventricular haemorrhagic venous infarction
  • Instability of cerebral blood flow (hypotension or fluctuations in blood flow)
  • Also some association with inflammatory exposure & endothelial injury

• **Risk factors**

• **Cardiovascular instability**
• **Hypotension**
• **Mechanical ventilation**
• **Pneumothorax**
• **Acute intravascular volume overload**
• **Hypercapnia**
• **Cerebral vasodilatation (direct effect)**
• **Chorioamnionitis**
• **Recognised association but more commonly related to PVL**
• **Endothelial injury**
Preterm infants (2)

- **Periventricular leukomalacia**
  - Injury to immature white matter prior to around 34 weeks gestation
  - Vulnerability of immature pre-oligodendrocytes to injury from various means
  - Pro-inflammatory cytokines - intrauterine or postnatal exposure
  - Oxidative injury (including free radicals from iron in extravasated blood)
  - Deep periventricular white matter is at the arterial borderzone of immature cerebral circulation

- **Risk factors**
  - Prolonged partial hypoxic ischaemia
  - Placental insufficiency
  - Intrauterine +/- intrapartum hypoxia / acidosis

- **Chorioamnionitis**
  - Exposure to pro-inflammatory cytokines
  - Mechanical ventilation
  - Instability of blood pressure secondary to intrathoracic pressure. Fluctuations in cerebral perfusion with impaired autoregulation leading to ischaemic injury

- **Hypocapnia**
  - Cerebral vasodilatation (direct effect)
  - Either severe hypocapnia (below around 2.6 KPa) or more prolonged exposure to moderate hypocapnia in a ventilated infant

- **Postnatal infection / NEC**
  - Related to systemic inflammatory response

- **Intracranial haemorrhage**
  - Iron in extravasated blood
  - Post-haemorrhagic hydrocephalus

- **Maternal ketonaemia**
  - Impairment of fetal oxygen delivery rather than absolute hypoxia
Neurological injury in preterm infants

• Causation often complex
  • Factors giving rise to preterm delivery itself
  • Chorioamnionitis
  • Potential prenatal sentinel event(s)

• Potential breaches of duty
  • Antenatal steroids
    • Reduction in risk & severity of RDS - relevant both for IVH & PVL
    • Reduction in risk & severity of IVH (primarily through this)
    • Meta-analysis describes less than 50 % reduction in risk & 24 hours prior to delivery
    • Physiological mechanisms are continuous - not a binary change
    • Arguments for significant contribution for either IVH/PVH or PVL with failure to administer antenatal steroids
  • Chorioamnionitis / exposure to intrauterine inflammation
    • Avoidance or duration of exposure
  • Hypocapnia in a ventilated infant
    • Profound hypocapnia or prolonged exposure to moderate hypocapnia
Hypoxic / ischaemic injury to the mature brain

Two principal patterns of injury

• Acute profound / near total hypoxic ischaemia
  • Injury to deep grey matter
  • Areas of highest metabolic demand in term infant
    – Thalami
    – Basal Ganglia
    – Perirolandic cortex
    – Brainstem

• Chronic partial / hypoxic ischaemia
  • Peripheral perfusion failure
  • Borderzone / watershed injury
Acute profound hypoxia / ischaemia

- Acute onset severe hypoxia / ischaemia
- Circulatory collapse
- Injury to areas of brain with highest metabolic requirements – beyond ten minutes
- Only in the event of survival following prolonged & aggressive resuscitation is there extensive injury in cerebral hemispheres
Acute profound hypoxia / ischaemia - conventional model

• Up to ten minutes non-damaging HI
• Circulatory collapse & injury to areas of highest metabolic demand
  • Ventrolateral thalami
  • Basal ganglia
  • Perirolandic cortex
  • Brainstem
• Duration beyond 25 minutes – death or very severe & extensive injury in survivors following very prolonged & aggressive resuscitation
Acute profound / near total HI
Clinical sequelae

- Typically dyskinetic cerebral palsy
  - Dystonia
  - Choreoathetosis
  - Bulbar problems

- Cognitive development may be relatively preserved in less severe cases
Timing of injury

- **Circulatory collapse & loss of perfusion**
  - Neurological injury within areas of highest metabolic demands
  - Consistent pattern of progression
- **The conventional “25 minute” model**
  - 10 minutes non-damaging
  - 15 minutes damaging HI
- **Variation from the “norm”**
  - Observed longer period of bradycardia
  - Prior hypoxia / acidosis
- **Damaging period is the terminal period of HI**
  - “Working forwards or backwards”
- **Resolved by restoration of circulation**
  - Variability likely to be within capacity to maintain critical level of perfusion
  - Some attempts made at 40:60 apportionment of observed bradycardia - no evidence base
- **Therapeutic hypothermia**
  - May have modulating effect on progression of injury (primarily basal ganglia)
  - Acute insult of mild to moderate severity most likely to benefit
Prolonged partial asphyxia

• Fetal hypoxia
  • Uterine hyperstimulation
  • Cord compression
  • Reduction of uteroplacental perfusion / placental insufficiency
  • Placental abruption
  • Fetomaternal haemorrhage

• Fetal heart rate decelerations (late / variable), reduced baseline variability +/- rising baseline correlating with fetal hypoxia & acidosis
Process of prolonged partial asphyxia

- Initially tolerated - non damaging hypoxic ischaemia (at least one hour, often considerably longer)
  - Redistribution of fetal blood flow
  - Increased cerebral blood flow
    - Cerebral oxygen delivery maintained despite hypoxia
- Increasing fetal hypoxia & acidosis leads to impaired myocardial function & perfusion
  - Loss of cerebral perfusion pressure & peripheral perfusion failure
- Hypoxic ischaemia at borders of cerebral circulation associated with swelling / oedema further compromising perfusion
- Infarction within arterial borderzones (watershed)
- Process does not progress in a linear fashion
  - Slow / arrested progression of injury, or
  - Rapidly escalating injury culminating in fetal circulatory collapse – “acute-on-chronic” picture
Neuropathological correlates

• Injury affecting the cerebral hemispheres
• Cortex / subcortical white matter
• Injury developing within the borderzones of distribution of major cerebral arteries
• Often diffuse cerebral oedema
Chronic partial / intermittent HI
Clinical sequelae

- Four limb spasticity
- Microcephaly
- Seizures
- Learning difficulties
- +/- sensory deficits
- Impairment of verbal IQ and cognitive impairment also reported in absence of motor disability
Timing of injury

• Onset of damaging hypoxic ischaemia
  • Often impossible to determine
  • Evidence of rapidly increasing / escalating HI makes it less likely that non damaging HI extends to greater than around 1 hour

• Continuing / escalating fetal hypoxia & acidosis
  • Intrapartum monitoring
  • Condition at birth
  • Cord gases
  • *Damaging* hypoxic ischaemia continuing until termination of the insult with continuing escalating hypoxia & acidosis

• Material contribution to neurological injury
  • Demonstration of increasing / escalating hypoxic ischaemia
  • Continuing neurological injury (final stages of insult damaging HI)
Additional factors in hypoxic-ischaemic brain injury

- **Blood glucose**
  - Hypoglycaemia
    - Increased risk for neurological injury with neonatal encephalopathy – increased corticospinal tract injury & worse cognitive scores
      *Tam et al, J Pediatr 2012;161:88-93*
    - Early hyper- and hypoglycaemia both correlated with adverse neurodevelopmental outcomes with neonatal encephalopathy
      *Basu et al, Arch Dis Child Fetal Neonatal Ed 2016;101:F149-55*

- **Infection / inflammation**
  - Sensitisation to hypoxic-ischaemic neurological injury
  - Biphasic effect dependent on timing of exposure
    - 4 – 6 hours & 72 hours
Resuscitation following perinatal asphyxia

- Reversal of hypoxia
- Restoration of perfusion
  - Lung inflation
  - Ventilation
  - Augmentation of circulation until recovery
    - Chest compressions
    - Epinephrine
- Supportive care
- Neuroprotection
Therapeutic cooling

- Neuroprotective measure
- Proven efficacy
- Well tolerated
- Mortality or major neurodevelopmental abnormality at 18 months
  - RR 0.75 (0.68 – 0.83), NNT 7 (5 – 10)
    
    *Jacobs et al, Cochrane Database of Systematic Reviews 2013*

- Survival with normal outcome
  - 1.53 (1.22 – 1.93), NNT 8 (5 – 17)
    
    *Edwards et al, BMJ 2010;340:c363*

- Toby-Xe (inhaled xenon as adjunctive treatment)
- No additional benefit demonstrated using MR spectroscopy
  
TOBY trial criteria

• Whole body cooling to 33 - 34°C
• Commence treatment within first 6 hours

Eligibility criteria:

• 36 weeks gestation or above
• 10 minute Apgar 5 or less or ongoing resuscitation
• pH <7.0 or BE $\geq$ 16.0 mmol/l in first hour

  and

• Moderate to severe encephalopathy
  – Clinical assessment
  – aEEG abnormal background $\geq$30 minutes or seizures

Azzopardi et al. NEJM 2009
Issues around cooling

- Decision to cool or withhold treatment
  - Application of accepted criteria
  - Failure to cool when indicated
  - Delay in initiating treatment
    - In light of evidence base, reduction in risk for neurological injury with cooling does not approach 50%
    - Extent to which cooling mitigates the severity of neurological injury in an individual case cannot be determined
- Effect of cooling on extent / severity of neurological injury
  - Impact on assessing likely timing of injury (with acute / profound hypoxic ischaemia)
Perinatal stroke

• **Arterial ischaemic stroke**
  • Focal injury due to critical ischaemia within specific arterial distribution
  • Most commonly embolisation from placental bed (intrauterine R to L shunting)
  • Rare without labour / uterine contractions
  • Carotid artery dissection / intimal injury
    • Extension / rotation & traction of head / neck at delivery

• **Risk factors**
  • Chorioamnionitis
  • Maternal diabetes
  • Pre-eclampsia
  • Conditions predisposing to thrombosis
  • Traumatic delivery (specific cases)

• **Deep cerebral venous sinus thrombosis**
  • Secondary associated intracerebral / intraventricular haemorrhage

• **Risk factors**
  • Infection (intrauterine or postnatal)
  • Observed association with perinatal asphyxia - causal mechanisms less clear
  • Conditions predisposing to thrombosis
Perinatal stroke - causation issues

• Arterial ischaemic stroke
  • Not a manifestation of global hypoxic ischaemia
  • Uncommon prior to labour
  • Discrete all or nothing event - although delay in delivery may increase duration of exposure to risk, cannot establish that arterial stroke due to embolisation more likely to have occurred prior to or subsequent to non-negligent time of delivery

• Deep cerebral venous sinus thrombosis
  • Secondary associated intracerebral / intraventricular haemorrhage
  • Risk factors
    • Infection (intrauterine or postnatal)
    • Observed association with perinatal hypoxia - no causal mechanism demonstrated
    • Conditions predisposing to thrombosis
    • Severe dehydration
Neonatal meningitis

• Most commonly bacterial meningitis
  • Bacteraemia
  • Bacterial proliferation
    • Septicaemia
      • Meningeal seeding of bacteria
      • Bacterial proliferation
        • Inflammatory response (meningitis)

• Bacterial colonisation / vertical transmission
  • Most common early onset infection Group B streptococcus (GBS)

• Clinical risk factors for infection
  • PROM
  • Maternal pyrexia
  • Clinical signs of chorioamnionitis
  • Known GBS colonisation

• Symptoms in newborn period
  • Lethargy
  • Poor feeding
  • Temperature instability
  • Cerebral irritation / seizures
Neurological injury with neonatal meningitis

• Most commonly following bacterial meningitis
• Focal infarction - endothelial injury & thrombosis in situ
  • Deep perforator artery
    – Basal ganglia, thalami & deep white matter
  • Small vessel ischaemia
    – Patchy focal cortical infarctions

  *Hernandez et al Pediatr Neurol 2011;44:282-8*

• Cerebritis & parenchymal injury
• Ventriculitis & hydrocephalus

• Hearing loss
Neonatal jaundice

• Bilirubin production exceeds capacity for conjugation & excretion
• Physiological
  - Mild unconjugated jaundice, self-limiting
  - Breast milk jaundice
• Pathological
  - Early onset (within first 24 hours)
  - Haemolysis
  - Excessive bruising
  - Infection
• Prolonged jaundice
  - Conjugated jaundice
  - Neonatal hepatitis
  - Cholestasis / biliary atresia
NICE neonatal jaundice guidelines 2010

• Evidence based consensus
• Visual assessment of jaundice unreliable
  • Any clinically apparent jaundice → bilirubin measurement (6 hours)
  • Bilirubinometer (<250 µmol/l) or formal serum bilirubin measurement
• Risk factors
  • Below 38 weeks gestation
  • Exclusive breast feeding
  • Previous sibling with neonatal jaundice
  • Ethnicity (Asian & African-Caribbean heritage – G6PD deficiency)
  • (Severe & extensive bruising)

• Hour-specific bilirubin measurements
  • Adapted to produce threshold table
## Hour-specific bilirubin

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<th>Bilirubin measurement (micromol/litre)</th>
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<th>Repeat bilirubin measurement in 6–12 hours</th>
<th>Consider phototherapy and repeat bilirubin measurement in 6 hours</th>
<th>Start phototherapy</th>
<th>Perform an exchange transfusion unless the bilirubin level falls below threshold while the treatment is being prepared</th>
</tr>
</thead>
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| **www.nice.org.uk**
Jaundice in preterm infants

• “Low bilirubin kernicterus”
  • Hypoalbuminaemia
  • Infection
  • Co-morbid white matter injury
    
    Watchko & Maisels Seminars in Perinatology 2014;38:397-406
  
  • Acidosis

• Importance of recognition of risk and trajectory of rise in serum bilirubin (rather than absolute serum bilirubin level)
Hypoglycaemic brain injury

• Cerebral neuroglycopenia
  • Symptoms contemporaneous with documented hypoglycaemia

• Neurological injury
  • (1) parieto-occipital cortex / subcortical white matter (2) more diffuse white matter injury consistent with hypoglycaemic injury

• Hypoxic-ischaemic injury
  • H-I injury exacerbated by hypoglycaemia: increased corticospinal tract injury, adverse motor & cognitive outcomes (Tam et al J Pediatr 2012;161:88-93)
Hypoglycaemic brain injury - causation issues

• Identification of high risk infants
  • Infant of diabetic mother (including gestational diabetes)
  • Intrapartum hypoxia & depletion of substrate +/- hyperinsulinism
  • Growth restricted infant (NB - customised antenatal growth charts)
  • Failure to establish feeding

• Inadequate observation & management
  • Early neonatal discharge prior to establishing effective breast feeding
  • Failure to support effective breast feeding (including offering appropriate supplemental feeds)
  • Failure to appropriately monitor feeding & blood glucose levels in at-risk infants
Questions
Have a nice Day!
Cerebral palsy - associated co-morbidities and complications.

Jill Cadwgan
Consultant Paediatrician, Neurodisability
September 2018
HANDOUTS FOR THIS PRESENTATION ARE CURRENTLY UNAVAILABLE AND WILL BE CIRCULATED SEPARATELY.
The Origins and Development of a Cerebral Palsy Integrated Pathway - CPIP

12th September 2018

With special thanks to Susan Quinn, Mark Gaston, James Robb, Laura Wiggins, Heather Read, Lesley Harper & Nicola Tennent
➢ **CP** - indicates that it is to do with cerebral palsy

➢ I – **integrated** - 'to combine two or more things to become more effective' (Cambridge dictionary)

➢ **P** - **pathway** to suggest that we are going or leading to something, with solid footing, showing the way
Cerebral Palsy

“An umbrella term covering a group of non progressive, but often changing, motor impairment syndromes secondary to lesions of the brain arising in the early stages of its development”
Cerebral Palsy

- Cerebral Palsy is a non–progressive neurological condition
- Progressive orthopaedic condition
- Associated impairments
- Reasons for hip displacement are multifactorial
- Children grow......
**What is Cerebral Palsy?**

Cerebral palsy is a physical disability that affects movement and posture. It is the most common physical disability in childhood.

**Motor Types**
- **Spastic:** 70-80%
  - Most common form
  - Muscles appear stiff and tight
  - Arises from motor cortex damage

- **Ataxic:** 6%
  - Characterized by shaky movements
  - Affects balance and sense of position
  - Arises from cerebellum damage

- **Dyskinetic:** 6%
  - Characterized by involuntary movements
  - Arises from basal ganglia damage

- **Mixed Types:** Combination damage

**Parts of the Body**
- Cerebral palsy can affect different parts of the body.

  - **Quadriplegia/Bilateral:** Both arms and legs are affected. The muscles of the trunk, face and mouth are often also affected.
  - **Diplegia/Bilateral:** Both legs are affected. The arms may be affected at a lesser extent.
  - **Hemiplegia/Unilateral:** One side of the body is affected.

**Gross Motor Skills**
- The gross motor skills of sitting and walking of children and young people with cerebral palsy can be categorized into 5 different levels using a tool called the Gross Motor Function Classification System (GMFCS) developed by CanChild in Canada.

**Manual Ability**
- At least two thirds of children with cerebral palsy will have movement difficulties affecting one or both arms. Almost every daily activity can be impacted.

**Associated Impairments**
- Children with cerebral palsy may also have a range of physical and cognitive impairments.

  - 1 in 3 is unable to walk
  - 1 in 4 is unable to talk
  - 3 in 4 experience pain
  - 1 in 4 has epilepsy
  - 1 in 4 has a behaviour disorder
  - 1 in 2 has an intellectual impairment
  - 1 in 10 has a vision impairment
  - 1 in 4 has bladder control problems
  - 1 in 5 has sleep disorder
  - 1 in 5 has saliva control problems

**World Cerebral Palsy Day**

[World Cerebral Palsy Day website](worldcpday.org)

Previously supported by The Arthritis Foundation

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[For more information, visit www.worldcpday.org]

[World Cerebral Palsy Day 2013]

[02 Oct 2013]
England, Wales & NI has a population of 58.8 million (53.9 England, 3.1 Wales, NI 1.8)

Incidence of 2-2.5 cases of CP per 1000 live births (not changed in 40 years)

Live birth rate approximately 695,000 in England & Wales giving an annual incidence of about 1500 new cases of CP per annum

Estimated total of 130,000 individuals with CP in England & Wales

Half of new cases will have severe CP (GMFCS 5) and an associated risk of hip displacement (Howard J, Soo B, Graham HK, Boyd RN, Reid S, Lanigan A, Wolfe R, Reddihough DS. Cerebral palsy in Victoria: Motor types, topography and gross motor function. J Paediatr Child Health 2005)
Meeting of Swedish Paediatric Orthopaedic Society & Scottish Paediatric Orthopaedic Club 2010
CPUP - Cerebral Palsy follow-up programme

- Began 1994 in southern Sweden
- Collaborative project between paediatric orthopaedic departments & community paediatric physiotherapists
- Recognised that a significant number of children with CP developed hip dislocation and severe contractures
- Aim was to prevent these complications through development of a system that would follow-up the child in a structured way throughout childhood
Orthopaedic Surgeons and children's physios meet in Perth 2011
Hip Displacement (MP>30%) & GMFCS

Soo et al., JBJS 2006
2800 children in Sweden who participate in CPUP
8 (0.02%) have dislocated
Majority of the 8 children were assessed as too seriously ill to undergo surgery.
Reduced number with severe contractures
Reduced number requiring surgery for scoliosis
“As so many are following-up children with CP using a standardised method, we have the unique opportunity to compare different treatment methods and drive forward research in CP within a bigger population. Adults with CP show improved function, less pain problem and reduced need for assistance and aids.”

Gunnar Hagglund
Common Goals of CPUP & CPIP

➢ A preventive follow-up programme for children with cerebral palsy or suspected cerebral palsy.

➢ To ensure that children, with risk of developing significant contractures or hip dislocation are detected early and receive treatment as early as possible.

➢ No child should be affected by severe contractures or hip dislocation and that every child should achieve the best function possible
Agreement that the knowledge and skills required to develop the surveillance programme and carry out the required measures were integral to physiotherapy practice – the skills required were “core business” for childrens physiotherapists

Agreed the need to standardise the measures used and ensure consistency with clinical assessments, resulting in a training need for contributing physiotherapists
CPIPS funded in 2013......
# Physical Examination

<table>
<thead>
<tr>
<th>Test</th>
<th>Range</th>
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<tbody>
<tr>
<td>Hip Abduction</td>
<td>&lt;20°</td>
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<tr>
<td>Popliteal Angle</td>
<td>&gt;60°</td>
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<tr>
<td>Knee Extension</td>
<td>&gt;10° fixed flex</td>
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<tr>
<td>Dorsiflexion/Knee flexed</td>
<td>&lt;0° (&gt;10°PF)</td>
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<td>Dorsiflexion/Knee extended</td>
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<tr>
<td>Internal Rotation</td>
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<td>External Rotation</td>
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<td>Ely test</td>
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<td>Hip Extension</td>
<td>&lt;10° (&gt;10°FD)</td>
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<td>Hip Extension</td>
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![Image of physical examination](image-url)
Cerebral Palsy Integrated Pathway Scotland (CPIPS). The project provides a central database for staff around Scotland for children with a diagnosis of cerebral palsy aged 2 and above. The aim is to provide a high quality, standardised follow-up programme for children with CP that will identify musculoskeletal problems by regular physical and radiological examinations to enable effective management of these problems during childhood.
Clinical Examination Details for GMFCS 1-3

Right True Leg length (mm) 63
Right Hip External Rotation (degrees) 50
Right Hip Internal Rotation (degrees) 80
Right Hip Flexion Deformity N
Right Hip Extension (degrees) 10
Right Hip Abduction Bilateral (degrees) 30
Right Hip Abduction Unilateral (degrees) 0
Right Hip RoM Pain (Y/N) N
Right Abduction hip neutral knee extended Fast (R1) (degrees) 0
Right Duncan Ely Test (degrees) 30
Right Knee Popliteal Angle (degrees) 90
Right Knee Fixed Flexion (degrees) 180
Right Knee Hyperextension (degrees) 90
Right Ankle Dorsiflexion knee flexed (degrees) 10
Right Ankle Dorsiflexion knee extended (degrees) 12
Right Ankle Dorsiflexion knee extended R1 (optional) (degrees) 35
Right FOOT WEIGHT BEARING Hind foot varus (Y/N) N
Right FOOT WEIGHT BEARING Hind foot normal valgus (Y/N) N
Right FOOT WEIGHT BEARING Hind foot excessive valgus (Y/N) Y
Right FOOT WEIGHT BEARING Midfoot break (Y/N) Y
SPINE Lumbar lordosis excessive (Y/N) N

CPIPS Assessment

SCOLIOSIS MANAGEMENT X-ray since last assessment (Y/N) N
SCOLIOSIS MANAGEMENT Surgery since last assessment (Y/N) N
Comments

SPINE Thoracic kyphosis excessive (Y/N) N
SPINE Scoliosis standing (Y/N) N

Generated on: 05/02/2014 20:47:39
(c) 2013 - University of Dundee
## X-ray Protocol

<table>
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<tr>
<th>Age</th>
<th>2</th>
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<tr>
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</table>

All children get a pelvic x-ray at two years, six years and at 16 years old before transition.

- Green annual x-ray
- Grey x-ray as necessary
- Yellow no x-ray
Standardised radiographic positioning
1. Legs parallel, patellae facing upwards.
2. Pelvis flat, lordosis reduced.
WHO: International Classification of Function (ICF)

Health Condition (e.g., CP, ASD)

Body Structure and Function

Activity

Participation

Environmental Factors

Personal Factors
## Functional Mobility scale

<table>
<thead>
<tr>
<th>Walking distance</th>
<th>Rating: select the number (from 1-6) which best describes current function</th>
</tr>
</thead>
<tbody>
<tr>
<td>5 metres (yards)</td>
<td></td>
</tr>
<tr>
<td>50 metres (yards)</td>
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</tr>
<tr>
<td>500 metres (yards)</td>
<td></td>
</tr>
</tbody>
</table>

**Rating C**

Crawling:
Child crawls for mobility at home (5m).

**Rating N**

N – does not apply:
For example, child does not complete the distance (500m).

**Rating 6**

Independent on all surfaces:
Does not use any walking aids or need any help from another person when walking over all surfaces including uneven ground, curbs, and in a crowded environment.

**Rating 5**

Independent on level surfaces:
Does not use walking aids or need help from another person. *Requires a rail for stairs. * If uses furniture, walls, fences, shop fronts for support, please use 4 as appropriate description.

**Rating 4**

Uses sticks (one or two):
Without help from another person.

**Rating 3**

Uses crutches:
Without help from another person.

**Rating 2**

Uses a walker or frame:
Without help from another person.

**Rating 1**

Uses wheelchair:
May stand for transfers, may do some stepping supported by another person or using a walker/frame.
**Figure 1:** Predicted Gross Motor Function Measure (GMFM-66) motor scores as a function of age by Gross Motor Function Classification level. *GMFCS levels with significant average peak and decline. Dashed lines illustrate age and score at peak GMFM-66.
Select CPIPS Report from list below:

1. Patient Summary Report
2. Patient Summary Report (XRay Median Means)
3. Patient Report (Red Zone XRays)
4. Examination Schedule Report
5. X-Ray Schedule Report
6. X-Ray Missing
7. Patient XRays
8. User Rights
CPIP UK
CPIP–UK Annual Meeting 2018
Sharing experiences of implementing CPIP and its impact on practice

A one-day event for paediatric physiotherapists, paediatricians and orthopaedic surgeons involved in the management of children and young people with Cerebral Palsy

Saturday, 3rd November 2018
Venue: Emirates Old Trafford, Manchester

The first CPIP-UK Annual Meeting will be held as part of the APCP Annual Conference

For further information / to book your place, go to: http://apcp.csp.org.uk/courses-events
Contact: courses@apcp.org.uk
Conclusion

With a cerebral palsy register, identifying all children with CP in a population, in combination with a screening programme it seems possible to prevent or reduce the development of severe contractures, hip dislocation and scoliosis in children with CP.

References


Independent Living Solutions Ltd.

Case Management and Rehabilitation Services
The Challenges of Paediatric Case Management

Anita Reals

Paediatric Case Manager and Occupational Therapist
• The role of the paediatric case manager
• Specific issues for children & families
• Case examples from ILS clients
Case Management is...

“...an active process devoted to the co-ordination, rehabilitation, care and support of people with complex, clinical needs and their families. It aims to facilitate their independence and improve their quality of life whilst acknowledging safety issues.”

British Association of Brain Injury Case Managers (2010)
‘A case manager is someone who will get on with the job, get their hands dirty, and make things happen.’

‘My right hand man...’
Who is your client?
Managing Expectations
Going at Family’s Pace

- Passing over care of your child
- Ready to accept equipment / adaptations
- Are parents ready to look to the future?
- Acceptance of their child’s disability
Robert
Matthew
Emotional Issues
‘It’s not so much the disability the person has but the person the disability has...’
William and George:

- 6 years old
- Severe cerebral palsy
- Gastrostomy fed
- Hoisted for all transfers
- Dependent on others for all care needs
- No verbal communication
- Live with their parents and two siblings
- Attended specialist schools
- Similar travel distances involved
Managing the Professionals

For George:

- 6 years old
- Severe cerebral palsy
- Gastrostomy fed
- Hoisted for all transfers
- Dependent on others for all care needs
- No verbal communication
- Attended specialist school
- Lived at home with parents and siblings
Educational Psychologist

Dietitian – local Hospital and Regional centre

GP, Health Visitor

Physiotherapist – CDC, School

Legal team

Speech and Language Therapist

Hospital – paediatrician neurologist

Occupational Therapist – NHS/School, Wheelchair service, Social services

Orthotics

Sensory impairment

School Staff, LSA, school nurse
Managing Care
4 years of case management costs for a child with severe cerebral palsy, aged 4 when CM intervention commenced
Impact on Siblings
How to motivates the Child or Young Person?
Key Messages

• Be aware of the impact of expert witness reports detailing a child’s long term needs on parents and the litigation process as a whole.

• Never underestimate the huge numbers of professionals (statutory, private, and legal) involved in these families lives and the impact of this on families.

• Time spent preparing families for having paid care in their home is essential.

• Ensure the case manager has a very clear picture of both the child’s needs and the family situation.
Contact Us…

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- CPD Training: Lindsay Oliver – External Training and Presentations Coordinator  
  T: 01722 742442  E: Lindsay.trainingpresentation@indliv.co.uk

- General Enquires  
  T: 01722 742442  E: mail@indliv.co.uk
Legal update on liability and quantum affecting CP cases

Henry Witcomb QC
Elizabeth-Anne Gumbel QC

Please see handout for notes

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0800 612 9660
Info.services@braininjurygroup.co.uk