Childhood hemiplegia

Questions to be answered

* What is meant by the term hemiplegia?
* How may it affect my child?
* Are there other hidden ways it may affect my child?

* What causes it?
* How is it diagnosed?

* What treatments are available
Hemiplegia is part of spectrum of cerebral palsy.
How is cerebral palsy defined?

**Group** of disorders affecting development of
- movement and posture
- permanent but not unchanging
- caused by a non-progressive disturbance in developing foetal or infant brain (up to 2 years)
- Can be accompanied by other problems e.g. with learning, perception, behaviour and epilepsy
Effects on development

* Motor development
  * Gross motor function- big movements e.g. crawling, walking
  * Fine motor function- small movements with hands e.g. manipulation, release of objects

* Communication (if dominant L hemisphere involved)
* Learning ability (Cognition)
Other possible effects on health & development

* Epilepsy- focal and generalised
* Visual problems- affecting visual fields- hemianopia
* Sensory problems- ignoring affected side - inattention
* Emotional and behavioural development
CP definition

* Defined by clinical description e.g. hemiplegia—one side of body affected due to damage to opposite side of brain
* Doesn’t tell you about what caused the damage or prognosis (future progress)
* Spectrum of disability – of movement problems and other difficulties too e.g. learning, communication, seizures, sensory impairments
* No two affected children exactly alike
Quadruplegia
All four limbs are involved.

Diplegia
All four limbs are involved. Both legs are more severely affected than the arms.

Hemiplegia
One side of the body is affected. The arm is usually more involved than the leg.

Triplegia
Three limbs are involved, usually both arms and a leg.

Monoplegia
Only one limb is affected, usually an arm.
Focuses on **movement** problems

* Sensory, learning & behavioural aspects not emphasised
* Variedness of disorder not made clear
* CP is not a specific diagnosis but **descriptive term for the umbrella of disorders**
* Comprehensive assessment of **whole child** needed because of range of problems
TYPES OF CEREBRAL PALSY

SPASTIC - tense, contracted muscles (most common type of CP).

ATHETOID - constant, uncontrolled motion of limbs, head, and eyes.

RIGIDITY - tight muscles that resist effort to make them move.

ATAXIC - poor sense of balance, often causing falls and stumbles.

TREMOR - uncontrollable shaking, interfering with coordination.
Commonest type of CP (25-40% of all cases)

* Antenatal cause in 75% cases, 10% post natal
* Involvement of arm and leg on one side (arm > leg). Least motor disabling
* Growth on affected side is poorer
* Silent interval before symptoms noted at 4-9 months - *early hand preference* commonest
* 67% are diagnosed by 18/12.
* > 50% affected children walk at average age
Spastic CP must have 2 of following:

- Abnormal pattern of movement and/or posture
- Increased tone - not necessarily all the time
- Abnormally brisk reflexes (Babinski)
- Spastic CP can be either unilateral or bilateral. **Unilateral is hemiplegia**
Parent usually first to suspect
- non use of one hand (hand dominance before 2 years is usually abnormal)
- Ignoring affected side
- Floppy or stiff limb
- Delay in acquiring motor milestone e.g. crawling, grasping item
Often nothing worrying around pregnancy or birth. “Out of the blue”

Or

* Prematurity, stay on neonatal unit & scans available from then
* History, examination and scans – all put together to make diagnosis
Plantar responses

- Normal plantar response
- Extensor plantar response (Babinski sign)
Components of classification

Doctor aims to include in report on child

* Motor problems
  * Tone- high/low/variable
  * type of movement problem- spasticity
  Associated problems e.g. epilepsy

* Anatomical & Scan findings
  * Parts of body affected
  * Neuro radiology scan findings

* Causation & timing
  * Identified cause- pre, peri or post natal
Unilateral spastic CP (Hemiplegia)

* No facial involvement (occurs in acquired hemiplegia)
* Intelligence- in normal range to slightly low
* Epilepsy occurs in $\frac{1}{4}$-1/3 of children with hemiplegia, often in those with learning and speech problems (often by 3 yrs) (Associated with cystic cortical lesions)
* Visual field defects occur in 25%
* Behavioural problems often greatest management problem
Physical effects of hemiplegia on arm

- Arm turned in at shoulder (pronated)
- Elbow bent (flexed)
- Held against side of body (adducted)
- Wrist bent (flexed)
- Fingers bent (fisted)
- Thumb held tightly in palm (adducted)
- Arm may look slightly smaller
Consequences of arm position, weakness and spasticity

* Difficulty with 2 handed tasks: DLA washing; dressing; feeding; toileting
* Play & School work: drawing; writing; computer work; cutting; using ruler; building; playing games
* PE: running; jumping; throwing and catching;
* Appearance & self confidence
* Independence
Mobility difficulties

- Toe walking (toe heel gait) - shortening of Achilles tendon (TA) and calf muscles
- Foot turned in (equinus) or out
- Knee bent and turned in (tight hamstrings)
- Hip flexed; limp
- Leg shortening; feels cooler; slimmer calf muscles
Consequences of leg weakness and spasticity

* Reduced stamina
  * Tires more easily
  * Complains of discomfort; spasms
* Poor balance
  * Needs hand rail on stairs
  * Unsure on uneven surfaces
  * Difficulty climbing; jumping; playing football
* Affects self confidence; inclusion by peers
* Physical appearance
<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>A: Hemiplegia</td>
<td>Arm, body, leg affected on one side</td>
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<tr>
<td></td>
<td>Arm turned in and bent</td>
</tr>
<tr>
<td></td>
<td>Hand fisted, Leg turned in and bent, Tiptoe standing.</td>
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<tr>
<td>B: Diplegia</td>
<td>Legs affected more than arms</td>
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<td></td>
<td>Arms slightly clumsy</td>
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<tr>
<td></td>
<td>Legs pressed together and turned in Tiptoe standing.</td>
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<tr>
<td>C: Quadriplegia</td>
<td>Whole body affected</td>
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<tr>
<td></td>
<td>Poor head control</td>
</tr>
<tr>
<td></td>
<td>Arms turned in &amp; bent</td>
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<tr>
<td></td>
<td>Legs pressed together Tiptoe standing</td>
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GMFCS E & R Descriptors and Illustrations for Children between their 6th and 12th birthday

**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.
Investigations/evaluation of child with suspected CP

* MRI scan- abnormal in 80-90% of all those scanned
* Helps with finding cause, prognosis but doesn’t alter management
* Clinical examination detailed history & MRI brain scan should determine cause in most cases
* Done at different times depending on how child presents but usually before 2 years
Prevalence of CP

- 2/1000 live births for all CP
- Half of all those affected were born pre term (≤32 weeks or ≤1.5 kg)
- 5% acquire CP after birth
- Hemiplegia is largest group within CP (29%)
- All CP more common in boys M:F ratio = 1.33:1
Causes of hemiplegia- perinatal stroke

Classified according to

1. **Ischaemic** (insufficient blood gets to area due to blockage of vessel or **haemorrhagic** due to breakage of vessel)

2. Blood vessel affected- artery or vein

3. Timing of injury- before or around time of birth (2/3 cases in utero)

4. Timing of symptoms- at birth with seizures or later in infancy with early hand preference
Left cerebral hemisphere showing specialised areas of cortex
Blood supply of important cortical areas
How do muscles receive messages from brain

* Message travels from cortex in cortico-spinal tracts
* crosses over in pyramids
* down into spinal cord
* emerges in biceps nerve in neck area
* passes to biceps muscle attached to upper arm
Cross section of brain to show motor tracts
Cortico spinal (pyramidal) tracts

Motor area of cortex

Internal capsule

Geniculate fibres

Decussation of pyramids

Anterior cerebrospinal fasciculus

Lateral cerebrospinal fasciculus

Anterior nerve roots
Stroke involving artery (at birth and antenatally)

This MRI picture of the brain arteries (MRA) shows an occlusion of the middle cerebral artery (MCA, arrow).

In NAIS, seizures in the newborn would lead to this MRI picture that shows an acute or “fresh” stroke.

In APPIS, an infant with weakness noted at 6 months would lead to this MRI which shows a loss of brain tissue in the same area.
Stroke involving veins occurring before birth

- Premature brain
- Veins (blue) drain the periventricular white matter

Germinal matrix bleeds -> Blocks draining veins -> Venous infarct (stroke) -> PVI damages motor pathways = weakness on opposite side
Management of movement problems

* Physical therapy - postural care, aiding development, stretching, hydrotherapy etc
* Orthotics
* Baclofen
* Surgery
Orthoses (splints)

* Muscle doubles length by 4
* Stretching is stimulus for growth
* AFOs
  * Restore heel toe gait
  * Stabilising joint
* Cumbersome
* Stretch gastrocnemius
* Little evidence from research
Botulinum Toxin A

* Inhibits acetylcholine release at nerve ending by temporarily blocking receptors at junction between nerve and muscle
* Muscles are relaxed as messages from nerve to contract muscle can’t pass across
* Botox or Dysport used
* Decreases spasticity, increases weakness

Botox also helps to reduce the appearance of wrinkles!
Botulinum Toxin A

* Useful in hemiplegia to reduce spasticity in lower limb to reduce
  * Equinus- abnormal foot down and turned in posture
  * Crouch gait due to knee flexion

In upper limb
* Elbow flexion (reach)
* Wrist/finger flexion
* Thumb in palm
* How to give injection: use sedation
  * Midazolam orally
  * EMLA topically
  * Gas and air to breathe in
  * Give injection under ultrasound guidance or by feel/site
* Review one month later
* Temporary improvement maybe beneficial
* Query long-term outcome
Set functional goals before starting. Help with

- Standing
- Walking
- Wearing splints
- Hygiene
- Pain/spasms
- Upper limb function- open hand, thumb out, wrist neutral position.
At around 7 – 9 years consider

* TA lengthening – after Botox finished

* Multi-level soft tissue release at
  * Hips - not usually required in hemiplegia
  * Knees
  * Ankles

* i.e. lots of small operations

* Rehabilitation so important
Other “hidden” problems

* Learning and attention problems may not be apparent until child starts school
* Awareness and recognition precede intervention
* Shouldn’t be attributed to other causes eg ‘naughty boy”
* Educational support needed
* May be exacerbated by other difficulties – epilepsy or visual problems
Other interventions

Behaviour

Often most prominent problem (occurs in 50%)
Sudden unexplained outbursts, may be associated with
* Aggressiveness
* Hyperactivity
* Epilepsy
* Delayed language development
* Learning problems
The future

- Emphasis on functional independence not correction of deformity
- Holistic approach & support for families
- Increased long term survival even in severe cases- need for more services
- New treatments- Botulinum, SDRR, IT baclofen infusion pumps
- Anti dystonia drugs
- Prevention of brain injury in neonates with neuro protective agents and cooling
Time for questions