ACPIN Conference 2014

Long-term implications of cerebral palsy in an aging population

Stephanie Cawker
Clinical Specialist Physiotherapist
Neuro-disabilities
Great Ormond Street Children’s Hospital

Stephanie.cawker@gosh.nhs.uk
Aims

Consider lifespan issues in people with cerebral palsy
GMFCS1-3

Current terminology & classification in CP

Legacy of childhood interventions and expectations

CP issues in adult management
Proposed definition of cerebral palsy

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

The motor disorders of CP are often accompanied by disturbances of sensation, cognition, communication, perception &/or behaviour &/or by a seizure disorder.

(Bax et al 2005)
1: 500 live births
Distribution and severity in cerebral palsy
Survey of CP in Europe

- Hemiplegia
- Diplegia
- Quadriplegia
- Dyskinesia
- Ataxia
- Other

Functional severity:
- Mild
- Moderate
- Severe
GMFCS Levels
Expanded and revised
Ages 12 - 18
Pallisano et al 2007
Predicted Average Development by GMFCS Levels

A = Can child lift head and maintain head in vertical position when supported at trunk

B = maintain sitting unsupported by his/her arms for 3 seconds

C = Walk unsupported 10 steps forward

D = Walking down 4 steps alternating feet with arms free

GMFCS I

- Independent community walker, uneven surfaces, and curbs
- Use stairs without using a rail
- Can run and jump
- Speed & balance are reduced
- Participate in sports
GMFCS II

- Walk in most settings
- Problems with long distances and uneven terrain
- Independent sitting
- May need help getting up & down from floor
- Stairs with rail
- Running & jumping limited
- May need adaptations to enable participation in sport
GMFCS III

- Walk using hand held device indoors
- Needs some support for sitting & walking
- Sit to stand transfers require assistance
- Use wheeled mobility for long distance
- Usually bilateral involvement with some UL problems
- Independent transfers but needs help to maintain alignment, base of support & foot clearance
FMS
The Functional Mobility Scale (version 2)

For children with cerebral palsy aged 4-18

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 etc. 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 the 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.

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

Rating C
Child crawls for mobility at home (5m).

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

www.rch.org.au
UMN Syndrome

Primary Injury

Loss of inhibition LMN

Positive features of UMN syndrome
- Spasticity
- Hyper-reflexia
- Clonus
- Co-contraction

Negative features of UMN syndrome
- Weakness
- Fatiguability
- Poor balance
- Sensory deficits

Musculoskeletal pathology
- Muscle shortening
- Bony torsion
- Joint instability
- Degenerative arthritis

Graham K
Primary Injury

UMN Syndrome

Loss of inhibition LMN
Positive features of UMN syndrome
- Spasticity
- Hyper-reflexia
- Clonus
- Co-contraction

Negative features of UMN syndrome
- Weakness
- Fatiguability
- Poor balance
- Sensory deficits

Musculoskeletal pathology
- Muscle shortening
- Bony torsion
- Joint instability
- Degenerative arthritis

CNS pathology
Loss of connections to LMN (and other pathways)

Neural
Mechanical
UMN Syndrome

Primary Injury

Loss of inhibition LMN

Positive features of UMN syndrome

- Spasticity
- Hyper-reflexia
- Clonus
- Co-contraction

Neural

Musculoskeletal problems

Muscle shortening

Bony torsion

Joint instability

Degenerative arthritis

CNS pathology

Loss of connections to LMN (and other pathways)

Negative features of UMN syndrome

- Weakness
- Fatiguability
- Poor balance
- Sensory deficits

Motivation

General health

Epilepsy

Intellectual & sensory deficits
Why is cerebral palsy different to UMN lesions in the adult population?
GROWTH
Brain, nerve, muscle and connective tissue
Maturity

Primary Injury
- Tone
- Balance
- Strength
- Selectivity

Secondary growth disorders
- Fixed contractures
- Mal-alignments and rotations
- Lever-arm dysfunctions

Coping

Tertiary compensations

Sensory motor learning
0-4 years

Activity, Function and participation
Adolescence

Growth
Cycle of Deformity - imbalance not spasticity
BUT Spasticity and growth accelerate deformity

Abnormal muscle activity and balance
Inefficient power

Muscle contracture
Joint integrity imbalance
Bony rotations
Lever arm dysfunction

Abnormal stresses lead to altered soft tissue metabolism & bone growth

Increased muscle, connective tissue and bone adaptation
Gait function and decline in adults with cerebral palsy: a systematic review

*Morgan et al* 2014

- 485 papers
- Mobility remains fairly stable from 15 to 25 yrs
- Unilateral CP adults continue to walk throughout adulthood but there are musculoskeletal issues
- Bilateral spastic CP GMFCS 3 (those using a wheelchair) decline earlier and are more likely to stop walking in late teens and 20s
- Dyskinetic CP are less predictable and are more likely to lose mobility due to MSK and balance deterioration
- In the older adult walking independently (GMFCS 1&2) at 60: 75% had dropped a level or died by 75
Walking into adulthood

Gains in mobility have been recorded right through to teens but high number of children who gain walking will lose it again or there will be a significant deterioration in capacity

Factors that contribute to whether adults continue walking can be complex and include environmental and personal factors

- Distance, location of work/school
- Energy demands – physical strain v work capacity
- Social preferences & personal interests

Sophia Werner
Paralympic athlete
What stops function in adulthood?
Clinical Prognostic Messages From a Systematic Review on Cerebral Palsy
Iona Novak, Monique Hines, Shona Goldsmith and Richard Barclay
*Pediatrics*; originally published online October 8, 2012;
DOI: 10.1542/peds.2012-0924
Pain - Prognostic messages

- Children and adults with contractures are at higher risk of developing pain.

- For those who can walk – neck, back and feet are high-risk pain sites.

- Pain increases with age.

- Pain is related to higher rates of behavioural problems and reduced levels of participation.

What’s causing their pain?

• Muscle and connective tissue pain

• Excessive overuse of distorted joints/alignment problems

• Joint deterioration - arthritis

• Injury, fractures – falls are common

• Previous or failed surgery
Hip displacement

Novak I et al Pediatrics 2012
Hip displacement

Novak I et al Pediatrics 2012
Hips and spine

- Children with both sides of the body affected and who cannot walk are at greatest risk of hip problems and scoliosis.

- The risk of hip abnormalities increases with physical disability.

**BUT:**
Over 1 in 10 children GMFCS II and 1 in 40 GMFCS III will also have hip displacement.

*Figure 1: Hip displacement (migration percentage >30%) by GMFCS Level (Soo et al. 2006)*
Hip subluxation: Varus osteotomy

Lumbar lordosis with Spondylololysthesisis

Crouch Gait  Knee flexion deformity + ankle equino-valgus

Can be a result of
calf weakness made worse by weight gain
over zealous calf muscle lengthening
lever arm dysfunction
hamstring and hip flexor tightness
Progression of patella alta and patellar fragmentation.

©2002 by Radiological Society of North America
Genu recurvatum.

More common in dyskinetic movement problems.


©2002 by Radiological Society of North America
Equinovalgus and rocker-bottom deformity.

Lever arm dysfunction
Energy inefficient

©2002 by Radiological Society of North America
In the *adducted/internally rotated shoulder*, the arm is held closely against the side, elbow bent, with the forearm applied across the front of the chest.

**Flexion of the wrist** is caused by hypertonicity of the wrist flexor muscles that seem to easily overpower their antagonists of wrist extension, so that this is the most common attitude.

The **flexed elbow** is bent into flexion and this posture may dramatically worsen with ambulation, causing more-severe angle flexion.
105 patients referred for orthopaedic problems
   Legs: shoe wear, pain and problems walking
   Arms: ADL and hygiene

92% had LL surgery
   including muscle lengthening & tendon transfers for flexible deformity
   arthrodesis and capsulectomy for fixed problems
   All who were ambulatory before surgery improved their status

40% had UL surgery
   Needed aggressive rehabilitation for functional gains
Movement disorder management

Similar to other adults with UMN lesions in terms of management options

NICE Spasticity Guideline 2012 for children and young people
Mentions dystonia

www.nice.org.uk/guidance/CG145
Muscle weakness

Secondary muscle pathology

- Spastic muscle has half number of sarcomeres in series but same fascicle length so reduced active force production *Matthewson m*
  
- Infiltration of collagen *Lorentzen*

- Muscle metabolism affected by abnormal activation *Smith L*

- Need to maintain muscle mass above critical threshold during adulthood to prevent premature sarcopenia *Shortland*

**The good news**

Muscle does respond to strengthening even in older adult population
Fitness and fatigue

Sedentary behaviour and deconditioning
- Posture deterioration
- Energy expenditure
- Muscular inactivity

*Verschuren et al 2014*
Need to increase MET across 24 hr period as well as more vigorous high intensity strength and fitness

‘Sit less move more’

‘Physiological burnout’
High energy costs & physical strain of activity that prevent capacity to function and not necessarily level of impairment
- Can’t work at optimum all the time
- Stress and fear
Painful Kyphosis and associated cervical hyperextension and chin poking

Sedentary lifestyle
More likely to spend working day sitting
Problems related to
- Poor posture
- Weak trunk muscles
- Reduced respiratory function
- Reduced hand function
- Associated problems
  - Poor use of vision
Bone health

- Genetic
- Nutritional
- Hormonal

- Environmental
  - activity
  - cultural

- Weight bearing with activity promotes mineralisation of bone

- NWB demineralisation osteopenia and potential risk of #

- # risk is most closely related to low body weight
The older adult with cerebral palsy >60

- Loss of nerve cells with reduced conductivity
  - can be greater in CP population and lower initial reserve
  - Decline in balance reduce response time and protective reflexes

- Muscle reduced speed of contraction and ability to repeat – fatigue
  - Can be greater due to previous surgery
  - Spastic muscles have fewer sarcomeres and increase extracellular matrix within the muscle - reduced optimal cross bridges to generate forces

- Effects of medication
  - Long term use of seizure meds can affect bone mineralisation

- Obesity, vascular disease and bone fragility (fracture)
  - Found to be similar to adults who acquire injury which leads to sedentary lifestyle

- Emotional and Psychological decline ? Isolation, care needs, loss of independence
Therapy Goals in CP management

*Moll et al*
Disability and rehabilitation 2013

Intensive physiotherapy as a child
‘Normalising Movement’

Early goals are mainly parent lead
Optimizing physical potential

In adolescence emphasis changes
Achieved full potential
‘no more to be done’
‘this is how you are’

Necessary compensations and environmental accommodations for optimal participation
Motor Management Options

**Early years ≤ 8 years**

**Physiotherapy and Occupational Therapy**
- Physical management programmes
- Direct interventions
- Handling and Postural management

**Equipment** for posture and function

**Splinting + Orthotics**

**Tone management**
- Botulinum Toxin A injections/Myoneural injections
- Oral drugs
- Selective dorsal rhizotomy SDR
- Intrathecal baclofen

**Orthopaedic Surgery**
- Hip surgery
- Muscle lengthening and bony realignment for severe problems

**Older child ≥ 8 years**

All those mentioned

Increasing use of Assistive technology

Multi-level muscle and bony surgery to correct alignment problems

Localised surgical procedures for instability, pain, correct alignment

Spinal and hip surgeries

Intrathecal Baclofen
Our challenge:

Improving life span care

How can we support adults to learn to use their bodies across their life course?

Think about the aging process and natural history of CP in order to preserve functional independence

Improve transition

Share learning within teams and at events like this!
Thoughts for the future

Selective dorsal rhizotomy

Functional electrical stimulation
Summary

• Advice on Lifestyle and Pacing
• Consider assisted mobility options /adaptations
• Regular low intensity activity as well as exercise for fitness
• Strengthening
• Maintain flexibility
• Address alignment issues - Orthopaedic surgery + postural management/Orthotics
• Spasticity movement disorder management
• Consider Emotional and Psychological health
Dedication to Mr Mark Paterson 1954-2013
Orthopaedic consultant RLHW
Thanks to my colleagues at GOSH neuro-disability service

stephanie.cawker@gosh.nhs.uk
www.nice.org.uk/guidance/CG145