Early Intervention – an update on the literature

Definition

Early intervention typically refers to a programme beginning within the first year of life for which the aim is to enhance infant development. The early years are critically important for cognitive and motor development. The timing of therapeutic approaches that support developmental acquisition during this period reflects the most dynamic period of neuroplasticity with the highest potential for ameliorating the negative sequelae associated with high-risk infants (Morgan, et al., 2016) (Hadders-Algra, et al., 2017).

Introduction

In infants, clinical signs and symptoms of cerebral palsy (CP) emerge and evolve before age 2 years. By 2 years was historically regarded as the latent or silent period where CP could not be identified accurately. Experts now consider the silent period as outdated because CP or ‘high risk of CP’ can be accurately predicted before 6 months corrected age. Early diagnosis should be the standard of care because contemporary early intervention and surveillance optimise neuroplasticity and prevent complications, therefore maximising functional outcomes, as well as efficient use of resources. (Novak, et al., 2017)

Early diagnosis of cerebral palsy

Half of all infants with CP have high risk indicators identifiable in the new-born period (e.g. prematurity, encephalopathy, seizures) enabling early screening. This population are described as having ‘new-born detectable risks for cerebral palsy’ and this pathway occurs before 5 months corrected age. For the other half of all infants with CP, the pregnancy and labour may have appeared to be uneventful and parents, caregivers or community-based professionals first notice delayed motor milestones. This population is described as having ‘infant detectable risks for cerebral palsy’ and this pathway occurs after 5 months corrected age (see Appendix 1 for Early Diagnosis pathway algorithm). (Novak, et al., 2017)

Alongside subjective clinical history taking to identify risk factors for CP, the most predictive objective tools for detecting risk before 5 months of age are neonatal magnetic resonance imaging (MRI), the Prechtl Qualitative Assessment of General Movements and Hammersmith Infant Neurological Examination (HINE). After 5 months corrected age, the most predictive tools for detecting risk are MRI, the HINE and the Developmental Assessment of Young Children. At any age, parental concern is a valid reason to trigger formal diagnostic investigations and referral to early intervention.

In terms of motor dysfunction, the infant’s quality of movement is reduced e.g. absent fidgety general movements (GMs) or neurological abnormalities (e.g. early observable hand function asymmetry or suboptimal HINE scores). In addition, the infant’s motor activities may be substantially below those expected for chronological age e.g. head lag, inability to grasp, not reaching for a toy when appropriate, inability to sit independently at 9 months or inability to weight bear. (Novak, et al., 2017)

To make an early clinical diagnosis this combination of assessments with strong predictive validity coupled with clinical reasoning is recommended. A highly experienced clinical team should ideally conduct and interpret the standardised assessments. Where the clinical diagnosis is suspected but cannot be made with any certainty, it is recommended to use the interim clinical diagnosis of ‘high risk of cerebral palsy’ until a diagnosis is confirmed.
Clinical diagnosis of CP or the interim diagnosis of high risk of CP should always be followed by a referral to CP-specific early intervention and surveillance protocol, this should include ongoing diagnostic monitoring where appropriate as well as standard medical investigations for associated impairments (e.g. hearing, vision, epilepsy). (Novak, et al., 2017)

**Hip surveillance**

There is a direct correlation between hip displacement and GMFCS level, and between hip displacement and type of CP, with increasing frequency in GMFCS III-V and in bilateral spastic CP types, with a strong trend towards hip displacement in non-ambulant children. (Terjesen, 2012)

Progression is from a normal hip at birth to a displaced hip as early as 2-3 years. (McClean, et al., 2014) In the Terjesen study hip dislocation was seen in children as young as 1yr 10m and mean age at dislocation was 4 years and 2 months. The likelihood of progressive hip displacement decreases with age, therefore it is under 5 years of age where hip surveillance is particularly key. (Terjesen, 2012)

Hip migration percentage (MP) is the percentage of the femoral head which sits outside the lateral margin of the acetabulum. It offers the most important single qualification of a hip joint. Hip displacement cannot be accurately diagnosed via active/passive range of movement assessment alone. (Terjesen, 2012). Hips are classified as normal under 33% hip displacement. Subluxed at MP 33-89% and dislocated at 90-100% MP. 33-39% MP represents ‘hips at risk’, subluxed hips can occasionally spontaneously resolve, though studies have found that the majority of hips in the range 50-60% MP range and beyond went on to dislocate. As an indication for preventive surgery 40% MP is generally recommended. (Terjesen, 2012)

Early, ongoing hip surveillance is required alongside timely surgical intervention in order to optimise longer term functional outcome, prevent the painful complications of hip displacement and maximise efficient use of resources. (Terjesen, 2012)

**Gross motor potential**

Early active movement and intervention are essential because infants who do not actively use their motor cortex risk losing cortical connections and dedicated function (Novak, et al., 2017). Children with CP reach 90% of their gross motor potential by age 5, with most potential achieved in the first 2 years. The first 2 years are regarded as a critical period for development of the corticospinal tract, therefore activity-based interventions during this period are vital for optimising outcomes. (Morgan, et al., 2016)

(Novak, et al., 2017) states ‘Randomised clinical trial data are beginning to indicate that 1) infants with hemiplegic CP who receive early Constraint Induced Movement Therapy (CIMT) develop better short and long term hand function 2) infants with bilateral CP who receive regular surveillance and intervention have lower rates of hip displacement, contracture and scoliosis 3) that infants with any type and topography of CP who receive Goals-Activity-Motor Enrichment (GAME), which is an early, intense, rich, task specific, training based intervention at home, have better motor and cognitive skills at 1 year than those who receive normal care 4) that improvements are even better when intervention occurs at home because children learn best in supported natural settings, personalised to their enjoyment’.

There is increasing evidence that the infant’s motor behaviour, via discovery and interaction with the environment, controls and generates the growth and development of muscle, ligament and bone, as well as driving ongoing development of the neuromotor system.
Physiotherapy and occupational therapy interventions should use child-initiated movement, task-specific practice, parent education and environmental adaptations that stimulate independent task performance because these interventions induce neuroplasticity and produce functional gains. These include Learning games curriculum (diplegia), CIMT or bimanual (hemiplegia), and GAME (all subtypes). (Novak, et al., 2017)

Tapping into neuroplasticity and maximising functional gains under age of 6-7 years is so important because we see that typical plateau followed by the possible decline in gross motor function, most commonly seen in GMFCS III-V. (Hanna, et al., 2009)

Therapeutic interventions for children with CP broadly encompass the breadth of the International Classification of Functioning, Disability and Health (ICF). Intervention aims to address body function/structure deficits, minimize activity limitations and improve functional skills, and encourage participation in age-appropriate settings or environment. (Morgan, et al., 2016)

![Figure 1: Predicted Gross Motor Function Measure (GMFM-66) motor scores as a function of age by Gross Motor Function Classification (GMFCS) level. *GMFCS levels with significant average peak and decline. Dashed lines illustrate age and score at peak GMFM-66. (Hanna, et al., 2009)](image)

International Classification of Functioning, Disability and Health Framework (World Health Organisation, 2007)
Severity and topography

Severity and topography are more difficult to ascertain in infancy as motor skills are still developing and presence or absence of hypertonia changes and evolves. Standardised tools (HINE, MRI) should be used to assess and monitor, and severity predictions should be made cautiously. Almost half of children younger than 2 have their Gross Motor Function Classification System (GMFCS) re-classified. Prognosis of long-term motor severity is most accurate in children older than 2 using the GMFCS.

In terms of motor subtype and topography, it is important to identify unilateral v’s bilateral CP because early intervention (e.g. constraint induced movement therapy), long term musculo-skeletal outcomes and surveillance needs differ e.g. likelihood of hip migration (See Appendix 2 – Clinical signs Indicating Motor Types and Topography in Infants). (Novak, et al., 2017)

Postural management

The Sunny Hill Health Centre for Children in Vancouver, Canada have developed a clinical resource tool based on research evidence and expert opinion that integrates GMFCS levels and ages from infancy to skeletal maturity to guide 24hr postural activity. It recommends that, whilst the role of positioning for prevention and management of hip displacement is emerging, early Intervention to promote motor development, arranging postural management equipment early and timely surgical intervention can avoid the painful and often costly complications of hip displacement. This includes potential impacts on care giving, participation, activity, sitting ability and sleep. (See Appendix 3 Positioning for hip health: a clinical resource) (McClean, et al., 2014)

With wind sweeping detectable from as early as 3 months corrected age, postural management should start in lying as soon as possible after birth (Gericke, 2006) and appropriately supportive seating systems should be in place from 6-9 months corrected age. Introducing a seating system and promoting upright posture can help avoid other areas of development suffering e.g. vision, communication and fine motor skills. (Paleg, et al., 2013).

Supported standing and weight-bearing should be introduced from 9-12 months corrected age and mobile weight-bearing should also be encouraged at an age appropriate stage (12-18 months). (Terjesen, 2012). Children with a motor disorder at GMFCS III require postural management programmes that emphasise postural activity from an early age (Gericke, 2006). For the appropriate children at GMFCS V a standing frame should be provided, for GMFCS IV both a standing frame and a gait trainer may likely be required. (Palisano, et al., 1997).

The recommended dose of weightbearing is 2 hours per day between standing frame and gait trainer. Children who ambulate less than 2 hours per day or are non-ambulatory often experience painful and costly complications because of extended periods spent in seated, supine, and prone postures. Supported standing programs have been used in various settings for more than 50 years in an effort to reduce and prevent complications and to optimise various aspects of function. Standing programs 5 days per week positively affects bone mineral density (60 to 90 min/day); hip stability (60 min/day in 30° to 60° of total bilateral hip abduction); range of motion of hip, knee, and ankle (45 to 60 min/day); and spasticity (30 to 45 min/day) (Paleg, et al., 2013)

Postural management equipment not only encourages improved alignment, preventing deformity, but also promotes function as well as facilitating peer interaction, activity and participation. (Terjesen, 2012)
Parents and caregivers

Parents and caregivers experience grief and loss at the time of diagnosis. They may mistakenly assume that the diagnosis means their child will need a wheelchair and have an intellectual disability. However, in high-income countries, population data indicate that 2 in 3 individuals with CP will walk, 3 in 4 will talk and 1 in 2 will have normal intelligence. 86% of parents of a child with CP suspect it before a clinical diagnosis is made. Parents and caregivers acknowledge that whilst receiving the information is difficult, they prefer to know earlier rather than later so that they can assist in their child’s development. Early detection helps the family foster acceptance and allows improved access to early intervention.

EI Smart (www.eismart.co.uk)

EI Smart is an approach being developed by a multi-disciplinary team of professionals within the UK. They recognise that whilst there is a wealth of research published on early intervention, there is no one approach bringing together the evidence-based components of effective interventions specifically for therapists. The evidence points to an approach where therapists acquire a broader skill set, shared across disciplines, including strategies for active engagement of the family and for supporting the family psychosocially.

The EI Smart approach proposes that the goal of early intervention is to minimise cognitive, motor and emotional impairments in young children disadvantaged by biological and environmental risk factors. The approach proposes consideration of the following core components:

1. Supporting a consistent and responsive parent-infant relationship
2. Challenging the infant with a wide variety of self-produced motor activities in a variety of conditions
3. ‘Scaffolding’ the infant’s next developmental steps
4. Minimising infant stress
5. Supporting the infant’s self-regulation
6. Promoting parental well-being

The EI Smart approach advocates a SMART start for high-risk infants, it recognises that one size does not fit all infants and whilst one intervention may be easier to evaluate, in clinical practice it may be better to adopt a pragmatic approach to intervention based on a combination of ingredients known to be effective in isolation.
SENSORY - the EI Smart approach recognises that sensory receptors are the interface between our CNS and the internal and external world. All motor behaviour, emotional and attention responses are as a result of how the brain processes sensory information. This processing creates behaviours that 1) keep us safe 2) drives us to explore and develop skills 3) regulates alertness levels/attention. Therefore, it is vital that an infant experiences an appropriate sensory environment to facilitate both neural and physical growth, promote exploration and assist development of self-regulation.

MOTOR - Infants who are developing typically begin life with high complexity and variability with redundant possibilities for postural control. Whilst learning new behaviours children explore many possible motor strategies, select a few strategies which are most efficient and reduce the use of the non-preferred strategies. Atypical variability is a key component in identifying motor control problems at an early age. Identifying atypical variability and enhancing the complexity of variability by encouraging a variety of infant directed motor actions and facilitating successful interactions via the ‘just right’ challenge may lead to functional changes and improvement in motor and cognitive function.

ATTENTION/ REGULATION – infants depend on caregivers to help them manage their emotional responses, in caring for infants parents are acting as extensions of their internal regulatory systems, over time infants develop their own strategies. Attention and regulation in early development is therefore deeply embedded in the child’s relationship with others, establishing the connection between parent and infant can be seen as the basic task of the early months of life.

RELATIONSHIPS - extensive research highlights the critical importance of mutually responsive interactions between parents and young children starting in infancy and its association with better developmental outcomes at preschool and school age. Therapists delivering the EI Smart approach aim to support a consistent and responsive parent-infant relationship, enabling parents to observe and interpret their child’s behaviours and modify the infant’s environment to promote high quality parent-infant interactions and parent-child synchrony.

THERAPY – the parent-therapist partnership is key to the successful delivery of EI Smart, supporting parents with the right information at the right time. Providing expert opinion communicated effectively to parents to facilitate them to deliver the ‘just-right’ challenge for their child. Involving the family in collaborative goal setting, ensuring awareness of support networks and driving positive expectation throughout.

Conclusion

Early detection of high risk infants, followed by specific and targeted early intervention, is recommended and should be the standard of care to optimise infant neuroplasticity, prevent complications, maximise efficient use of resources and enhance parent and care-giver well-being.

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May 2018
Appendix 1

Figure. Algorithm for Early Diagnosis of Cerebral Palsy or High Risk of Cerebral Palsy

Newborn detectable risks
- Premature
- Encephalopathy
- History of neurological risk factors (e.g., birth defect, IUGR)
- Parent identified concern
- Unable to sit at 5 mos or hand asymmetry

Infant detectable risks

Risk or concerns warrant an investigation for CP

Conduct a medical history and clinical examination with or without investigations for etiology and differential diagnosis (as indicated)

If <5 mos CA

A

Clinical neurological examination
- 4.1 HINE

Neurological imaging
- 3.2 MRI

Motor tests
- 3.1 CAMc
- 3.2 TIMP

Combined assessment data indicates

- 1.1 High risk of CP
- 1.2 Uncertain

If >5 mos CA

A

Clinical neurological examination
- 6.1 HINE

Neurological imaging
- 6.2 MRI

Motor tests
- 6.3 CAMc
- 6.4 TIMP

Combined assessment data indicates

- 1.1 Uncertain
- 1.2 Definitely MCI/CP

Determine preliminary severity of CP
- 8.1 HINE >53
- 8.1 MRI WMI
- 8.1 HINE <47
- 8.1 MRI GMl

Likely ambulant
Likely nonambulant

Determine preliminary topography

Assess for associated impairments

Communicate findings to parents compassionately

Arrange early intervention and parent support

Monitor

Confirm diagnosis

A indicates the best available evidence pathway. B indicates the next best available evidence pathway when some pathway A tools are not available. The numerals correspond to the numbering in Table 1. AIMS indicates Alberta Infant Motor Scale; CA, corrected age; CP, cerebral palsy; DAYC, Developmental Assessment of Young Children; GMs, Prechtl Qualitative Assessment of General Motor Movements; HINE, Hammermith Infant Neurological Examination; IUGR, intrauterine growth restriction; MAs, Motor Assessment of Infants; MFl, magnetic resonance imaging; NMDA, Neuro Sensory Motor Development Assessment; TIMP, Test of Infant Motor Performance; and WMI, white matter injury.
Appendix 2

Table 2. Clinical Signs Indicating Motor Type and Topography in Infants

<table>
<thead>
<tr>
<th>Unilateral Spastic Hemiplegia</th>
<th>Bilateral Spastic Diplegia</th>
<th>Bilateral Spastic Quadriplegia</th>
<th>Dystonia</th>
<th>Ataxia</th>
</tr>
</thead>
<tbody>
<tr>
<td>GM1</td>
<td>Cramped synchronized GMs, followed by 90% absence of EMG activity</td>
<td>Cramped synchronized GMs, followed by 90% absence of EMG activity</td>
<td>Early onset and long duration of cramp-like GMs, followed by 90% absence of EMG activity</td>
<td>Poor repertoire GMs, followed by absent fidgety movements and finger spreading</td>
</tr>
<tr>
<td>MRI</td>
<td>Fetal vascular insults (24%)</td>
<td>Malformations (3%)</td>
<td>Bilateral hemisphere (grade II) with or without hypoplasia</td>
<td>Lesions in the parietal white matter involving the thalamus</td>
</tr>
<tr>
<td>HINE Score15</td>
<td>≤50</td>
<td>≤50</td>
<td>≤50</td>
<td>≤50</td>
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<tr>
<td>50-73</td>
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<tr>
<td>Motor Tests</td>
<td>Asymmetrical hand preference</td>
<td>Good hand function compared with lower limbs</td>
<td>Head lag</td>
<td>Twisting arm or neck posture on voluntary movement (may be painful)</td>
</tr>
<tr>
<td></td>
<td>Stiff or floppy posture (50-73)</td>
<td>Persistent tardy function compared with lower limbs</td>
<td>Flexed posture in supported sitting</td>
<td>Head lag</td>
</tr>
<tr>
<td></td>
<td>Causal or avoidance of floor sitting</td>
<td>Weight bears on one leg or foot</td>
<td>Bilateral fisted hands</td>
<td>Slow to reach and grasp with either hand</td>
</tr>
<tr>
<td></td>
<td>Reduced variation in motor behavior</td>
<td>Reduced variation in motor behavior</td>
<td>Reduced variation in motor behavior</td>
<td>Reduced variation in motor behavior</td>
</tr>
</tbody>
</table>

Abbreviations: CP, cerebral palsy; GMFCS, Gross Motor Function Classification System; GPE, PEDI Qualitative Assessment of General Movements; HINE, Hamersham Infant Neurological Examination; MRI, magnetic resonance imaging; PLIC, posterior limb of internal capsule; PVE, periventricular echogenicity; PVL, periventricular leukomalacia.
Appendix 3

Positioning for Hip Health: A Clinical Resource
Sunny Hill Health Centre for Children, Vancouver, BC, Canada

Introduction
The development of hip displacement and dislocation in children with cerebral palsy (CP) is well documented in the literature [1, 2]. The progressio is from a normal hip at birth to a displaced hip as early as 2-3 years [3]. Hip displacement/dislocation and its resulting pain can have a huge negative impact on care-giving, participation, activity, sitting ability and sleep [4, 5]. Management of hip displacement/dislocation primarily involves medication and surgery. Clinically, positioning in lying, sitting, standing and walking is used pre and post hip surgery to address many goals such as participation and activity, self-care, sleeping and comfort. The rise of positioning for prevention and management of hip displacement and/or dislocation is emerging. The objective of this poster is to illustrate a clinical resource tool that integrates GMFCS levels, age, from infancy to skeletal maturity and positioning interventions. The resource is based on research evidence and expert opinion.

Research Evidence
As a clinical team we reviewed the evidence of positioning as a treatment approach for the prevention of hip displacement and/or dislocation. Our research question was:

Among children with CP does “positioning” affect the hip development?

The research evidence is insufficient to demonstrate a strong causation between positioning and hip development. This is due to the relatively low numbers of studies, the mostly moderate and weak quality of the studies and the low level of the study designs. The “traffic light” code and state of the evidence classifications is yellow [6].

The traffic light designation of yellow is symptomatic, indicating insufficient evidence and an urgent need to measure outcomes.

Péter References

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