IC 31: Pain in Cerebral Palsy across the lifespan: An evidence-based approach to understanding, assessing, and treating complex pain.

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In the following presentation, our frame of reference will be that of the bio-psycho-social model launched by Engels in 1977 (1) and further developed into the WHO’s International Classification of Functions (ICF) framework for classification of health and function (2).

Fig 1: International Classification of Function (WHO)

In the ICF, pain is allocated to the dimension of body functions, chapter 2 “Sensory functions and pain.” Cerebral palsy (CP) is the health condition defined as “a group of permanent disorders of the development of movement and posture, causing activity limitations that are attributed to non-progressive disturbances in the developing foetal or infant brain. The motor disorders of CP are often accompanied by disturbances of sensation, cognition, communication, perception, and behaviour, in addition to epilepsy and secondary musculoskeletal problems.” According to the report, the musculoskeletal problems include pain (3).

1: Pain in children and adolescents with cerebral Palsy (Kjersti Ramstad)

The systematic literature review published by Illona Novak in 2012 describes our target population: “Among children with cerebral palsy, 3 in 4 were in pain; 1 in 2 had an intellectual disability; 1 in 3 could not walk; 1 in 3 had a hip displacement; 1 in 4 could not talk; 1 in 4 had epilepsy; 1 in 4 had a behaviour disorder; 1 in 4 had bladder control problems; 1 in 5 had a sleep disorder; 1 in 5 dribbled; 1 in 10 were blind; 1 in 15 were tube-fed; and 1 in 25 were deaf”(4).
The IASP definition of pain was launched in their Journal “Pain” in 1979 and is much in use. It tells that pain is “an unpleasant sensory and emotional experience associated with actual or potential tissue damage, or described in terms of such damage.” The usual definition of chronic pain is pain lasting more than 3 months.

Pain in CP includes the experiences of people across the whole lifespan, people with or without intellectual impairment, and it may be reported by the person in pain him(her)self or by a proxy; usually a caregiver (parent or professional). The “gold standard” for pain report in paediatric CP is self-report whenever possible in addition to proxy-report.

In CP, the disease-specific target organ is musculoskeletal tissue. Potential sources for stimulation of nociceptors are muscle overuse, strain caused by involuntary movements, immobilization and atypical joint compression from the imbalance of muscle activation across joints. The term musculoskeletal pain refers to anatomy and actual or potential damage of musculoskeletal tissue. It is a somewhat ambiguous term that relies on clinical handles. The character of musculoskeletal pain is said to be dull or aching (as opposed to terms like burning and sharp).

Early literature on pain in paediatric CP describes a variety of orthopaedic issues. Newer literature tends to include complex concepts such as Quality of Life (QoL) and Health Related Quality of Life (HRQL) in which bodily discomfort or pain is included. Fatigue is a concept more seldom used in the paediatric literature than in reports on adults with CP. The literature on associations between pain in paediatric CP and participation may be labelled flourishing. Still, the literature on a common problem in a relatively common medical condition is far from overwhelming. A search in PubMed July 2015 for “pain AND cerebral palsy AND child” revealed 502 hits. Endpoints of the studies may be sorted out in two groups: 1) Musculoskeletal issues and 2) HRQL, QoL, Fatigue, Mental Health and Participation.

The “Index” study of pain in paediatric CP is the SPARCLE study initiated from Newcastle, UK by Alan Colver and his co-workers. Children included in European CP registers were visited in their homes at the age of 8-12 years (SPARCLE I) and revisited four years after (SPARCLE II). The SPARCLE homepage gives study design, summaries and scientific papers.

Study of Participation of Children with Cerebral Palsy Living in Europe
http://research.ncl.ac.uk/sparcle/
The paper “Self-reported quality of life of 8-12-year-old children with cerebral palsy: a cross-sectional European study” was published in the Lancet (5) and is much commented and referred to. From the abstract:

METHODS: 1174 children aged 8-12 years were randomly selected from eight population-based registers of children with cerebral palsy in six European countries and 743 (63%) agreed to participate; one further region recruited 75 children from multiple sources. Researchers visited these 818 children. 318 (39%) with severe intellectual impairment could not self-report; 500 (61%) reported their QoL using KIDSCREEN

FINDINGS: Impairments were not significantly associated with six KIDSCREEN domains. …. Pain was common and associated with lower QoL on all domains. Impairments and pain explained up to 3% and 7%, respectively, of variation in QoL.

In another paper, parental proxy-report on child QoL is compared to children’s self-report. The main findings are described like this in the abstract (6): ”We found that high levels of stress in parenting negatively influenced parents’ perception of their child’s quality of life, whereas the main factor explaining parents’ ratings of children’s quality of life higher than the children themselves is self-reported severe child pain.”

Further, parental proxy-report on child QoL was compared to the proxy-report given by professional care-givers (7). Parental stress and child pain again influenced the reports: “High levels of stress in parenting negatively influenced parent reports of child QoL compared with professional reports, while child pain was associated with professionals rating lower than parents.”

The estimated population prevalence of self-reported pain in the previous week was 60% in SPARCLE I and 74% in SPARCLE II. For parent-reported pain, the estimated prevalence was 73% in SPARCLE I (last 4 weeks) and 77% in SPARCLE II (last week) (8,9).

In SPARCLE I self-reports, older children reported more pain but pain was not significantly associated with severity of impairment. In parent reports, severity of child impairment, seizures and parental unemployment were associated with more frequent and severe pain (9).

In SPARCLE II, results are summarized like this (8):

About 75% experience some pain in a typical week.
40% experienced leg pains
34% reported headaches
45% of those who received physiotherapy experienced pain during therapy.

Girls reported more pain than boys (OR=2.1, 95% CI 1.5 to 3.0)

Young people reported more pain if they had emotional difficulties (comparing highest and lowest quartiles: OR=3.1, 95% CI 1.7 to 5.6).

Parents reported more pain in children with emotional difficulties (OR=4.2, 95% CI 2.7 to 6.6), or with more impaired walking ability.
We now leave the SPARCLE study and take a look at a hospital based study from Canada (10). One-quarter of the sample (N=252, age 3-19 y) experienced pain that limited activities and participation. Physicians identified hip dislocation/subluxation, dystonia, and constipation as the most frequent causes of pain.

A study by Barney et al is restricted to musculoskeletal pain (11) and also differentiate at various aspects of pain. Findings were that GMFCS level significantly predicted parent-reported musculoskeletal pain frequency (P<0.02), duration (P=0.05) and intensity (P<0.01) (N=34, mean age 9.4y).

In Oslo, we studied recurrent musculoskeletal pain (RMP) in a convenience sample of children and adolescents 8-18 years old. We found that RMP was the dominating pain and that age above 14 years predicted RMP (N=153) (12). Contrary to the SPARCLE study, we obtained self-report on mental health. We found that girls, but not boys with RMP self-reported a higher level of peer problems (p=0.02) than peers without RMP (13).

Summing up:

| Children and adolescents with CP experience more pain than typically developing youth |
| Children with severe CP experience more pain than children with less severe CP (parental report) |
| Musculoskeletal pain is the main pain problem |
| Gender differences emerge during adolescence |
| Caregiver and child report on pain are not identical |

Notes:
Treatment to reduce pain

In the treatment of pain in CP, complex interventions targeting more than one ICF dimension is the rule. As evidence is still scarce, we will address both empirical based best practise and user knowledge in addition to research based knowledge.

The national Cerebral palsy Follow-up Program in Norway (CPOP), which is a motor follow-up program, registers among several other parameters the prevalence of pain in all children with CP. CPOP shows that 92% of the children have physiotherapy, 64% 1-2 times a week or more. The prevalence of pain is 29%, increasing with lower functional level. However, only 10% get physiotherapy targeting pain (Annual report 2014).

4. Physical Therapy treatment for pain in persons with CP (Reidun Jahnsen)

One Norwegian study (Jahnsen et al 2003) describes the experiences of physical therapy and physical activity reported by adults with CP, both what they remembered from their childhood and current experiences. The treatment targeting pain will be reported.

Physiotherapy and Physical Activity – Experiences of Adults with Cerebral Palsy, with Implications for Children

Objectives: To investigate self-reported experiences, needs, type and amount of physiotherapy and physical activity in adults with CP, the variables associated with regular physical activity
and physiotherapy in adulthood, and the potential contribution of physical activity and physiotherapy in relation to prevention of chronic pain, fatigue and deterioration of function.  

*Design:* A postal survey on adults with CP in Norway.  

*Results:* The respondents were 406 persons with CP, without intellectual disabilities, 49% females, 18-72 years (mean 34 y). Type, amount and experiences of physiotherapy and physical activity were reported in addition to socio-demographic data, subdiagnosis, functional level, and data on new health problems, such as pain, fatigue and deterioration. Almost all the respondents (92%) received physiotherapy as children, while 40% had physiotherapy and 46% performed physical activity regularly as adults. The most common type of treatment was exercise on a bench (83%), stretching (78%) and walking exercise (58%). Women were nearly 50% more likely to use physiotherapy than men. Persons at GMFCS 3–5 were four times more likely to use physiotherapy than those at GMFCS 1–2. One third experienced pain from the physiotherapy as children and 6% as adults. However, 50% reported pain relief from physiotherapy as adults. Those who had learnt the meaning of physical activity as a child were three times more likely to be physically active as adults.  

*Interpretation:* The study documents that physical activity may contribute to prevent deterioration, chronic pain and fatigue in adults with CP, however, functional level was the strongest predictor. The strongest predictive variable significantly associated with regular physical activity as adult, was having learnt how to take personal responsibility for personal health during childhood. These experiences give valuable information, which could be used to make better life span follow up programs for the persons with CP.

A recent study by Slaman et al (2015) describes a lifestyle intervention program for adults with CP.  

*A lifestyle intervention improves fatigue, mental health and social support among adolescents and young adults with cerebral palsy: focus on mediating effects*  

*Objective:* To evaluate the effect of a lifestyle intervention on fatigue, participation, quality of life, gross motor functioning, motivation, self-efficacy and social support, and to explore mediating effects of physical behaviour and physical fitness.  

*Design:* A randomized controlled trial with intention to treat analysis.  

*Setting:* Rehabilitation centres in university hospitals in the Netherlands.  

*Subjects:* Adolescents and young adults with spastic cerebral palsy.  

*Interventions:* A six-month lifestyle intervention that consisted of physical fitness training combined with counselling sessions focused on physical behaviour and sports participation.  

*Main measures:* Fatigue, social participation, quality of life and gross motor functioning.  

*Results:* The lifestyle intervention was effective in decreasing fatigue severity during the intervention (difference = –6.72, \( p = 0.02 \)) and in increasing health-related quality of life with respect to bodily pain (difference = 15.14, \( p = 0.01 \)) and mental health (difference = 8.80, \( p = 0.03 \)) during follow-up. Furthermore, the domain participation and involvement of the social support increased during both the intervention (difference = 5.38, \( p = 0.04 \)) and follow-up (difference = 4.52, \( p = 0.03 \)) period. Physical behaviour or physical fitness explained the observed effects for 22.6%, 9.7% and 28.1% of improvements on fatigue, bodily pain and mental health, but had little effect on social support (2.6%).  

*Interpretation:* Fatigue, bodily pain, mental health and social support can be improved using a lifestyle intervention among adolescents and young adults with cerebral palsy. Furthermore, substantial mediating effects were found for physical behaviour and physical fitness on fatigue, bodily pain and mental health.
Vogtle et al (2014) have conducted a recent study on an exercise program for pain and fatigue in adults with CP.

**Outcomes of an exercise program for pain and fatigue management in adults with cerebral palsy.**

**Purpose:** The purpose of this exploratory pilot study was to examine the effect of exercise on pain and fatigue in adults with CP.

**Method:** Twenty-six participants (12 ambulatory, 14 non-ambulatory; 10 males, 16 females; mean age 42.3 SD 11.2 years) enrolled in a study using a repeated measures design including baseline, intervention and follow-up phases of 12 weeks each; 20 participants completed all phases. Primary outcome measures used were the FACES pain scale, the count of body parts with pain and the PedsQL Multidimensional Fatigue Scale.

**Results:** Significant beneficial changes were found in the pain and fatigue scales among the ambulatory participants during the intervention phase. However, the beneficial changes diminished during the follow-up phase. Secondary outcomes examined included, pain interference, daily physical activity and health-related quality of life.

**Conclusion:** Study outcomes suggest that exercise may provide some benefit for ambulatory adults with CP.

Riquelme et al (2015) describes procedural pain related to physiotherapy in persons with CP.

**Are physiotherapists reliable proxies for the recognition of pain in individuals with cerebral palsy? A cross sectional study.**

**Background:** Pain is an important problem for individuals with cerebral palsy (CP). In addition to pain associated to the pathology, individuals with CP are often exposed to physiotherapy procedures which may cause or relieve pain.

**Objective:** The major aim of this study was to compare pain ratings self-reported by individuals with CP and ratings about pain in others provided by their physiotherapists.

**Method:** Cross-sectional study. Children and young adults with cerebral palsy (n=50) and their physiotherapists (n=18) completed semi-structured interviews about clinical pain, as well as about procedural pain and pain relief elicited by standardized health procedures. Moreover, pain ratings were obtained during the application of hamstring stretching and passive joint mobilization.

**Results:** Moderate-to-high agreement was observed between individuals with CP and their physiotherapists on presence and intensity of pain, pain interference with physical activities and current and retrospective pain ratings elicited by physiotherapy procedures. By contrast, agreement regarding pain relief elicited by physiotherapy techniques was low.

**Conclusions:** Our data suggest that although physiotherapists may be reliable proxies for the recognition of pain in individuals with cerebral palsy, further research should be done to improve the communication between health professionals and individuals with CP around pain.

Hirsh AT et al (2011) have investigated pain treatment and self-reported effects in a survey on adults with CP.

**Survey Results of Pain Treatments in Adults with Cerebral Palsy.**

**Objectives:** To identify the types and frequencies of pain treatments used by individuals with cerebral palsy (CP); examine the perceived effectiveness of these treatments; and identify the types of healthcare providers that were accessed for pain-related services.

**Design:** A cross-sectional survey design was employed and 83 adults (mean age=40.3 years, SD=13.6) with CP participated.
Results: 63% of participants reported experiencing chronic pain and rated their pain intensity over the past week as 5.1/10, on average. The most common pain locations were the lower back, hips, and legs. Physical interventions (e.g., physical therapy, strengthening) were the most common pain treatments reportedly used, and were rated as moderately effective. Many other treatments were also used, and participants sought pain-related care from a variety of providers.

Conclusions: Although participants reportedly accessed pain care from a variety of providers, and perceived that several types of treatments were effective, many of the treatments rated as effective were rarely used or provided. Future research using clinical trial methods would further elucidate the specific pain treatments that are most beneficial for adults with CP.

5: Treatment of pain in children and adolescents (Kjersti Ramstad)

Implications of the SPARCLE I study (From the SPARCLE Homepage):

- Children should be asked whether they have pain by their parents and professionals. Perhaps a child, thinking their pain was “normal” because he/she had always had pain, does not mention it.
- Treatments should be developed for pain – the common headaches and tummy pains through psychological approaches.
- Pain due to cerebral palsy itself needs trials of medication or other treatments to deal satisfactorily with it.
- Some treatments for cerebral palsy may actually cause pain. These treatments should be carefully reconsidered to make sure they do really help the children.

Lessons from the SPARCLE II study according to the paper of Parkinson et al (8):

- About 75% experience some pain in a typical week. Strategies to reduce pain appear to be absent or inadequate.
- Clinicians should routinely assess pain and develop pain management plans if necessary.
- Much pain is caused by clinical procedures or therapies; their efficacy should be re-examined to establish if they deliver sufficient benefit to justify the pain and fear of pain that accompany them.
The assumed cause of pain should of course guide the treatment.

<table>
<thead>
<tr>
<th>Causal treatment</th>
<th>Treatment of the motor disorder</th>
<th>Treatment aimed to keep or restore body alignment</th>
</tr>
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<tbody>
<tr>
<td>dental caries</td>
<td>spasticity</td>
<td>Positioning, orthoses</td>
</tr>
<tr>
<td>gastroesophageal reflux</td>
<td>dyskinesia</td>
<td>PT</td>
</tr>
<tr>
<td>constipation</td>
<td></td>
<td>orthopaedic surgery</td>
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<td>malfitting orthoses</td>
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The conditions in the first column are all well known in CP, and have to be ruled out or well treated before treatment of recurrent musculoskeletal pain is negotiated. Regarding the topics in the second and third column the endpoints treatment of pain and prevention of pain (both at present and in the future) seems to blur both in clinical practise and in the literature. A multidisciplinary group in UK has developed guidelines for treatment of spasticity in children for the National Institute of Health and Care Excellence (NICE clinical guideline 145/guidance.nice.org.uk/cg145) issued in July 2012 (14) called “Management of spasticity and co-existing motor disorders and their early musculoskeletal Complications.” Their initial notions on delivery of care are excellent and may as well be applied to pain management plans. The guidelines discuss the following treatment options of which only oral drugs can be said not to cause pain:

Oral drugs: diazepam, baclofen, trihexphenidyl, levodopa
Botulinumtoxin-A
Intrathecal baclofen
Orthopaedic surgery
Selective Dorsal Rhizotomy

What do families do themselves to relieve pain in the child with CP?
Evidence is scarce or lacking. In our study of RMP, rest and change of position were the main pain relievers. 33 out of 95 children with RMP had taken an over-the-counter drug such as paracetamol or ibuprofen to relieve RMP during the last four weeks. Three children with RMP did not take any measures to relieve pain (12). A study from UK on alternative or complementary treatment (CT) in children with moderate or severe CP points to pain as a primary reason to try CT and also addresses the main challenge of understanding which factors predict optimal outcomes in CP (15).

When does RMP become chronic pain? The term chronic pain is not much elaborated in the paediatric literature on CP. On the other hand, mechanisms in and treatment approaches to paediatric chronic unexplained pain are well described. A web search for “Explain Pain” may be useful.

Summing up:

We are beyond the time when pain in paediatric CP was not recognized
We face much uncertainty about best practice of treatment
We should be protective of our special interest in RMP; both spontaneous and procedural
Our frameworks for understanding and treatment should be based on a bio-psychosocial approach to health, family-centeredness and include the life-span perspective
References:


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