Pain Assessment in Children with Cognitive Disabilities: Critical Review and Clinical Implications

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Abstract

Effective pain management is widely acknowledged as a core determinant for the provision of high quality care for both acute and chronic paediatric health care settings. Children with cognitive disabilities (CD), including intellectual disabilities (IDs), and cognitive impairments, have been identified as being at greater risk of experiencing pain and commonly they are lacking the ability to communicate pain adequately. Scope of this special article is to point out the importance of self-reporting with the use of simple pain tools in these children and to provide readers with a tool set for observational and proxy assessment. Moreover, introduces the idea of a systematic mixed method of assessment for adequate pain recognition in every setting. The main barriers for pain assessment in children with cognitive impairment are verbal restrictions and their inability to communicate pain as well as the atypical way they commonly express pain. Therefore, the use of an individualized approach with a mixed method of self-report, proxy report, and observational validated tools is highly recommended. The development of specific tools designed to detect and assess pain in children with intellectual disabilities is an important step in the assessment and management of the pain they experience. Since there is no ideal tool to achieve an accurate measurement of pain, the most appropriate evaluation scale according to the circumstances, or a combination of different tools, should be implemented. Clinical and translational research should focus more on the development, validation and reliability of pain assessment tools for children with cognitive impairment.

Keywords: pain assessment, pain management, pain scales, cognitive disabilities, cognitive impairment

Introduction

Adequate pain management is widely acknowledged as a core determinant for the provision of high quality care for both acute and chronic paediatric health care settings. Pain is a complex, subjective and stressful experience that is usually perceived through self-report (Breau et al, 2003). Children with cognitive disabilities (CD), including intellectual disabilities (IDs), and cognitive impairments, have been identified as

being at greater risk of experiencing pain, lacking the ability to communicate pain or even having limited cognitive capacity to understand what is happening to them (Clarke, 2015). There is evidence that this paediatric population often receive inadequate pain management and consequently adequate pain assessment is essential for their safety and comfort (Cascella et al, 2018; Best, Asaro & Curley, 2019). Additionally, compared to typically developing children, children with developmental (cognitive) disabilities were twice as likely to visit the emergency room, four times more likely to be hospitalized (Newacheck, 2004).

Research regarding pain in children with CD has emerged the last five years since in the past, due to their verbal abilities, they were excluded from most studies where self-report was considered a gold-standard. The children with even mild CD may not be able to fully express their experienced pain, due to limited or no verbal communication, shifting the responsibility of detecting pain to the caregiver or the health professional who must decipher and assess the behavior and signs in order to decide whether these signs are related to pain or not. Moreover, the children with CD usually present pain in an atypical way, which may lead to difficulty distinguishing between symptoms of the psychopathological disorder and those suggestive of pain (Einfeld et al., 2006).

Given the particular individual cognitive and communication abilities of children with cognitive disabilities and the consequent difficulties in assessing pain, the rate of undertreatment of pain in children who are unable to self-report it is increasing (Chen-Lim,et al, 2012). In all these cases of undertreatment, the inability to communicate pain is the main problem. This lack, in fact, can further confuse the observer, who mistakenly believes that the child does not perceive any pain. (Cascella & Muzio 2017).

Given high hospitalization rates and the prevalence of cognitive impairments in children, is essential the use of reliable & validated pediatric pain measurement tools, appropriate for the level of the mental development of children with CD and depending on whether or not they can self-report pain, since children with these disabilities cannot report pain verbally, but are able to report pain by communicating it with i.e. adaptive devices, symbols (Crosta, et al.2014) (Dubois,et al, 2010).

Since the 1990s, numerous pain measures have been developed and used in children with CD. These pain measures contain similar content and focus on pain behaviors, but vary in form, parent or caregiver involvement, psychometric properties, and clinical utility, creating challenges for clinicians who are called upon to determine the most appropriate measure to use in an acute care setting (Crosta, et al,2014). However, the studies including children with CD are limited with a great number of them emerging the last five years. Consequently, various characteristic-specific instruments suitable for specific conditions have been proposed, such as the Face, Legs, Activity, Cry, Consolability scale (FLACC), the Pediatric Pain Profile (PPP), the Children's Non-Communicating Pain Checklist (NCCPC) and the INRS Individualized Numeric Rating Scales, aimed at this specific patient population.

Assessment of pain in children with Cognitive Disabilities

In children with cognitive disabilities, pain is a significant problem due to the many and complex medical conditions and the numerous painful medical procedures they often undergo. The assessment and management of pain in these children is often complicated by communication barriers, which make inadequate the management of this issue, the treatment of which is a primary goal of the therapeutic approach (Breau et al, 2003). The difficulty in investigating pain symptoms is that assessment can be very difficult in these cases. Although pain assessment tools exist, they are typically used in children without cognitive disabilities and, therefore, may not be reliable for use in these specific conditions. (Crosta et al,2014).

Several self-reported & observational pain assessment tools have been used in children with CD. Self-reported measures are considered of paramount importance for pain assessment. However, their use is depending on the severity of CD that affects the tool's reliability. Therefore, children with borderline or mild to moderate cognitive impairment should be assessed with a self-report tool. Based on the current evidence Faces Pain Scale has been successfully used in children with severe CD. In comparison to numerical scales, Faces Pain Scale has greater performance even in children with severe CDs (Fanurik et al, 1998). In general, children with CD use simplified scales better (Dubois, Capdevila,

Bringuier, & Pry, 2010; Zabalia, 2013). However, health professional prior to their use should assess children's ability to fully comprehend and ability to use the selected self-report scales (Cascella et al, 2018). On the other hand, children with insufficient communication skills and severe cognitive disabilities, observational pain assessment tools may be more appropriate. Given that there is no ideal measurement tool for this population, an accurate assessment and evaluation of pain can often be achieved by using the most appropriate tool, or by combining different tools.

Assessment of pain in non-communicative intellectually disabled children, or in children with severe cognitive disabilities, is assessed indirectly by observing physiological changes, such as breathing, skin color, sweating, or through verbal expressions such as aggressive behavior, and body posture or movement. Standardized tools have been developed including sets of potential pain indices. The most used are based solely on behavioral responses or a combination of physiological changes and behavioral responses. A very effective strategy, adopted by various observational tools, involves parents in the process of assessing their child's pain. Parents have been shown to be more familiar with their child's normal behavior than clinicians, who usually have no previous experience with the child. However, proxy reports have limitations based on the current evidence. For example, mothers become adept at assessing their child's pain without input from health professionals, although there is concern that parents may tend to overestimate children's symptoms, particularly in situations of acute pain in emergency settings, or tend to underestimate it when the disease is chronic, mild or not fully verbalized (Carter et al, 2002; Matziou et al, 2016).

Reliable and validated observational scales for the assessment of pain in children, applied according to the child's age and ability to communicate with the caregiver, in different clinical settings (Merkel, et al., 1997). Each tool has specific characteristics and is suitable for application under specific conditions. The most used tools are those with established validity and reliability in children aged 3-18 years who cannot self-report pain due to cognitive impairment and conditions such as cerebral palsy, congenital or chromosomal syndromes, autism, seizure disorders, neurodegenerative diseases and encephalopathy.

In the following section the most commonly used tools are briefly presented.

The original and revised Face, Legs, Activity, Cry, Consolability scale (FLACC) scores and the revised FLACC (rFLACC;

The original and revised FLACC scores were developed as a simple observational tool aimed at assessing pain in children who are unable to verbalize the presence or severity of pain. This tool includes five categories of pain behaviors: facial expression, leg movement, activity, crying, and comforting. Each category is scored on a scale of 0-2 (Table 1).The total score ranges from 0 to 10. The cumulative score is classified as follows: mild (0–3), moderate (4–6) or severe pain (7–10). When children are awake, they should be observed for at least 1–2 minutes, while the examiner should observe the uncovered legs and body, assessing tension and tone, and intervening to comfort the child if necessary. Sleeping children should also be examined for at least 2 minutes. Several validation studies have been conducted to verify the reliability of this scale in assessing pain after surgery, trauma, cancer, or other disease in preterm children in which the original FLACC represents the most commonly used tool (Manworren, & Hynan 2003;Voepel-Lewis et al, 2020; Twycross, & Collis, 2013). Surveys into the potential implementation of the tool in cognitively impaired children undergoing surgical procedures, showed that facial expressions were the most sensitive indicators of pain compared to motor behaviors (Terstegen et al. 2003). Therefore, the original tool was further adapted by expanding the behavioral descriptors and leaving space for the description of individualized behavior.

The revised FLACC (r-FLACC) is a suitable tool for the assessment of acute pain in children with special needs including children with severe neurological impairments such as cerebral palsy (Malviya, et al,2006). It is easy to use even without the presence of parents(Chen-Lim, 2012), and this makes it applicable during hospitalization, especially in acute care settings (Ely et al, 2012). The tool (r-FLACC) considers the comfort factor and has significant reliability and validity in the assessment of pain after malignancy, surgery, medical procedures, trauma, and other diseases. The inclusion of an open-ended descriptor to incorporate individual pain behaviors, makes the revised version suitable for children with atypical pain behaviors that could

not be identified through other pain assessment tools (Cascella & Muzio, 2017).

Table. 1. Criteria for the FLACC pain scale

Behavior	0	1	2
Face	No particular expression or smile	Occasional grimace or frown, withdrawn, disinterested	Frequent to constant quivering chin, clenched jow
Legs	Normal position or relaxed	Uneasy, restless, tense	Kicking or legs drawn up
Activity	Lying quietly, normal position, moves easily	Squirming, shifting, back and forth, tense	Arched, rigid or jerking
cry	No cry (awake or asleep)	Moans or whimpers; occasional complaint	Crying steadily, screams, sobs, frequent complaints
Consolability	Content, relaxed	Reassured by touching, hugging or being talked to, distractible	Difficult to console or comfort

The Pediatric Pain Profile (PPP)

The PPP is designed to assess and monitor behavioral pain in children with severe neurological impairment. Includes child's pain history, baseline and ongoing pain assessments, interventions and outcomes, and discussion with clinicians about child's pain. It consists of a 20-item rating scale. Each item is rated on a four-point ordinal scale ranging from 0 (not at all) to 3 (very much) at any given time. The total score (PPP score) ranges from 0 to 60. A PPP score in the range of 10–19 is associated with "mild pain", 20–29 with "moderate pain" and 30 or more with "severe pain" (Hunt et al, 2004). In general, scores greater than 14 indicate moderate to severe pain, although this may vary from child to child. Parents or caregivers first complete the tool to determine baseline scores on a "good day," as well as when the child is experiencing pain. These scores become the benchmark against which it follows,

and scores are compared. A 5-minute observation period is recommended before scoring.

This scale uses a series of criteria covering the five key dimensions of PPP purpose and yields a project score. The user of the screening tool is asked to define the criteria related to each dimension on which the projects are evaluated. Each criterion is then broken down into a set of underlying indicators linked to a simple scoring system. The value of the criterion is calculated by summing the values of its indicators. To determine the value of an attribute, the result of each criterion is aggregated and finally, the values of all five dimensions are summed to arrive at the project score, which is calculated for each project and then plotted to demonstrate its suitability for PPP in the short term, medium or long term (or not at all). The relevance of the results depends on the stage of the project and the availability of information. The result of a PPP screening exercise is a shortlist of projects that can be considered "first movers"

for PPP development. PPP has been shown to correlate strongly with global pain ratings (performed by independent researchers via video analysis) and to be effective in children with "high pain" and "low pain" (Hunt et al, 2004; Hunt et al, 2007).

Furthermore, it is a useful tool for children with severe disabilities (e.g. vision, hearing or communication), and due to the possibility of monitoring pain and treatment effectiveness, it could be useful for children with chronic or recurring pain (Breau & Burkitt, 2009). It is easy to use by trained observers/parents (takes no more than 5 minutes to complete the assessment). It is also suitable for implementation in the assessment of postoperative pain. In addition, this tool includes self-injurious behaviors, which represent a common type of destructive behavior in children with a greater degree of cognitive impairment (Summers, et al, 2017). Compared to the r-FLACC, parents found the PPP more accurate although was considered less easy to be administered (Chen-Lim et al, 2012).

However, the PPP is not suitable for diagnosing pain, as it is designed to describe and record pain-indicating behaviors, monitor pain and the effectiveness of treatments, and facilitate communication between parents and caregivers about child's pain. Since this tool is designed to be used systematically by parents, skill development in the correct use of the scale is essential. Furthermore, this scale cannot be used effectively in certain clinical scenarios such as acute conditions or after surgery, due to the length of the tool (Hunt, 2011).

Pain Behaviour Checklist (PBC)

The Pain Behavior Checklist was developed to assess the frequency of four dimensions of pain behaviors, such as distorted wandering, emotional distress, facial/auditory expressions, and help seeking (Kerns, et al, 1991). These four dimensions comprise a set of 17 pain behaviors that were derived from a pool of 63 hypothetical "pain behaviors" (Turk, et al., 1985). The initial version was followed by the development of the 23-item PBC to assess postoperative pain in children with profound cognitive impairment (Terstegen et al., 2003). Of these 23 items, only 10 (mainly regarding signs in facial expression) can be divided between "absence of pain" and "presence of pain". The reduced PBC is another version of the PBC that consists of 10 nonverbal

items such as tears, panic attacks, soft cries, sad/near tears, grimacing, moaning, deepened nasolabial fold, eyes closed, tense face, and each of these items is scored positive or negative if the behavior is observed for 2 seconds or more (Duivenvoorden, et al., 2006). The PBC score ranges from 0 to 10 if all items are rated positively. A study of children who suffered from a combination of severe intellectual and motor disabilities (PIMD) showed that pain detection was more effective in these children. An important advantage of the 10-item version is the ability to distinguish between "absence of pain" and "presence of pain", and it can also be applied to cases of acute pain and profound pain. Although the original version of the tool is more accurate, it requires a lot of observation time, opposite to the simplified ten-item version which requires less time, however it may be less accurate than the original version for certain behavioral items (e.g., squinting) (Duivenvoorden, et al., 2006).

The Non-communicating Children's Pain Checklist (NCCPC- PV)

The NCCPC is a 31-item pain checklist designed specifically for children with cognitive disabilities who cannot communicate verbally (McGrath et al, 1998). The Pain Checklist for Noncommunicative Children-Revised (NCCPC-R) was designed by Breau et al. (2002), to assess pain during daily life and is intended for use by parents or caregivers who typically care for the child with a disability, at home or in a long-term care facility. The NCCP-r is structured into seven categories: vocal, social, facial, activity, body, and limb, physiological, and nutritional/sleep. The interpreter of the NCCPC-r must answer the question: "How often has the child shown these behaviors in the last 2 hours?", scoring 0 to 3, 0 = not at all during the observation period, 1 = a little, 2 = quite often, and 3 = very often/almost constantly. A total score greater than 7 indicates that the child feels pain, and this score has been confirmed in 84% of cases. There is also the postoperative version of the tool (NCCPC-PV)55 which explores 27 types of behaviors in six subscales, scored from 0 (not observed at all) to 3 (observed very often) during a 10-minute observation. Compared to the original version, the NCCPC-PV excludes items related to eating/sleeping. The composite score can range from 0 to 81. A score of 11 or greater can detect 88% of children with clinically significant pain, while a score of 6–10 detects mild pain with 75% accuracy (Breau,et al,2002). The NCCPC-r was

designed for untrained parents and caregivers but can also be used by adults unfamiliar with the child. Another important advantage of the tool is that the pain assessment in the NCCPC-r is not affected by the child's developmental level (Breau,et al,2011).

The Non-Communicating Children's Pain Checklist – Postoperative Version (NCCPC-PV), validated by Breau et al. (2002), is a tool for clinical evaluation and measurement of pain in children aged 3 to 18 years, with intellectual disabilities who cannot communicate. This instrument, intended for use by physicians, was designed to assess postoperative pain in noncommunicative children, specifically to assess pain after surgery or during procedures performed in the hospital. The NCCPC-PV scale is a parent or clinician report of observed behaviors, consisting of 27 items. Each item is scored from 0 to 3 based on six subscales (ie, vocal, social, face, activity, body, and limb, normal). It yields a total pain score ranging from 0 to 81. A score between 6 and 10 indicates mild pain, while scores > 10 indicate moderate to severe pain. The NCCPC-PV requires the caregiver or clinician to observe the child for 10 minutes before assigning the pain score. The NCCPC-PV could be useful in the diagnosis of pain, has high interobserver reliability, and familiarization with the child is not considered necessary (Breau, et al., 2002), however, the length of this checklist and the wide range of scoring could make it difficult to apply as a routine pain assessment in clinical settings (Ghai,et al, 2008; Cascella M, Muzio,2017).

Individualized Numeric Rating Scaleς (INRS)

The INRS Individual Numerical Rating Scale is based solely on individual pain indices by defining pain intensity through parent reports of their children's unique pain behaviors. Health care professionals interview parents and/or caregivers to describe their children's pain behaviors. Once described, responses are then ranked on a scale ranging from 0 to 10 (0 = no pain, 5 = moderate pain, and 10 = worst possible pain) (Solodiuk et al., 2010). The INRS is created individually for each patient according to the interpretations and reports of parents or caregivers about the child's pain behaviors. Although the instructions do not specify the required observation time, an observation time of one minute was used to validate the scale. INRS versus NCCPC-PV (comparison study) after surgery, based on ratings

provided by parents, nurses, and research assistants, found moderate correlations between INRS and NCCPC-PV and a significant decrease in INRS scores one hour after a pain management intervention. In the study, there was strong inter-rater reliability, however, bedside nurses consistently scored lower pain scores compared to parents and nurses. Although parents reported that they were able to complete the assessment without difficulty, for their children without difficulty, the time required to complete it was not measured to provide a full picture of the clinical utility of the INRS (Solodiuk et al., 2010).

The INRS is an adaptation of the numerical rating scale that incorporates parents' (and/or caregivers') descriptions of their child's past and current responses to pain. The INRS was originally developed at Boston Children's Hospital to help critical care nurses observe, consistently document, and communicate between shifts, their unique pain markers in children following major surgery. In April 2000, hospital-wide practice guidelines recommended the use of INRS in all clinical areas (Solodiuk, 2010).

Clinical Implications

Paediatric nurses should in their daily practice have the adequate skills and knowledge to manage pain in in hospitalized children. Especially for children with CD that are unable to self-rate pain intensity. The use of mixed methods that interfere observational tools, the task of selecting case by case the most appropriate pain assessment tool, and the use of proxy reports, are highly recommended. Furthermore, in the frame of family centered care model they should discuss with parents and about how the child expresses pain based on the child's cognitive and communication abilities. Scope of this multidimensional assessment approach is to improve patient outcomes and relief pain.

Self-reported measures are considered of paramount importance for pain assessment. However, their use is depending on the severity of CD that affects the tool's reliability. In daily practice, nurses should be able to distinguish which children with CD are able to self-report pain intensity. Therefore, the use of simple self-report pain tools is recommended. In addition, health professionals should be aware that children with CDs may not be able to verbalize their pain and that this may lead to under-treated and under-recognized pain.

The power of personal beliefs and culture should also be recognized. Such examples include the perceptions that children with CDs have elevated pain thresholds or that any child can verbalize pain intensity. It has to be clearly stated and acknowledged that pain is a common condition experienced by children with CD. Emerging evidence suggests that children with CD may be more sensitive to painful stimuli, face consequences from untreated pain and are more likely to experience chronic pain (Cascella et al, 2018; Best, Asaro & Curley, 2019).

Nowadays, as already presented in the previous section of this article, there is a great number of pain assessment tools that can been used in children with CD, independently of the severity of their cognitive impairment. However, further research on their reliability and validity is needed.

Another important point that we need to point out for daily clinical practice is that children with cognitive impairments may exhibit idiosyncratic and typical pain behaviors when they do not have pain, making it difficult for health professionals to discern signs of pain. Therefore, the parents' pain assessment and a well established communication with them is essential. Health professionals and parents as observers may not be sufficiently sensitive to the magnitude of pain the children with CD experiences. There is evidence that observers systematically underestimate or overestimate patients' suffering due to their stereotyped beliefs, and this is especially crucial in children with CD that self-report is not available or reliable. Therefore, more research in the field is required in order to support the use of valid and reliable measures specific to pain in children with CD. As well as translational research must focus on their implications in everyday clinical practice. Knowledge transfer into practice is also affected by organizational factors, including organizational culture and structure (Wysong, 2014).

The key message that should be taken from this critical review article is that children with CD commonly express their pain in an atypical way and the health professionals should focus more in a systematic approach for its assessment and management with the use of individualized approaches and validated assessment tools. Painful conditions are common in this vulnerable group (Breau & Camfield, 2011).

Conclusions

In children with cognitive disabilities, pain is an important issue because of the multiple and complex medical problems in these children. Pain assessment and management are often complicated because of many communication barriers, which make it extremely difficult to assess, interpret, and effectively manage pain in this population. The assessment of pain in children in this population is very complex and difficult, mainly to the verbal restrictions and their inability to communicate pain as well as the atypical way they commonly express pain. Therefore, the use of an individualized approach with a mixed method of self-report, proxy report, and observational validated tools is highly recommended. The development of specific tools designed to detect and assess pain in children with intellectual disabilities is an important step in the assessment and management of the pain they experience. Since there is no ideal tool to achieve an accurate measurement of pain, the most appropriate evaluation scale according to the circumstances, or a combination of different tools, should be used.

Despite these challenges, healthcare providers are professionally and ethically obligated to ensure competent individualized pain assessments for children with CD. Clinical and translational research should focus more on the development, validation and reliability of pain assessment tools for children with cognitive impairment. Moreover, health professionals need to invest in their training to acquire the knowledge and skills to use adequately pain scales. And in a holistic approach we should remember that parents are valuable partners in conducting a complete pain assessment beyond the quantification of pain and successfully manage not only pain intensity but the consequences from the whole pain experience.

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