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Assessing and Treating Pediatric Pain and Developmental Disabilities

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ASSESSING & TREATING PEDIATRIC PAIN AND DEVELOPMENTAL DISABILITIES

INTRODUCTION

Most research specifically addressing the pain of children with cognitive impairments has emerged within the past decade. This group has been included in few studies of pain in children in the general population because most incorporate some aspect of self-report, which is not always available from this group, or because it is assumed their pain experience or response will differ from typical children. They are also typically excluded from studies of specific painful conditions, because they often have several medical conditions that may complicate assessment and/or management of pain. Children with cognitive impairments form a heterogeneous group. Those with mild intellectual impairments may have no visible physical impairments and can communicate verbally. In contrast, those with the most severe intellectual impairments frequently have severe physical limitations and may neither speak nor understand spoken language to any practical extent. Children with cognitive impairments are more likely to be blind or deaf, and many display autistic behaviors. This has led some to believe their pain behavior is erratic, and cannot be used to reliably judge their pain. However, this also means that pain assessment tools developed for typical children may not be suitable for this group.

The exclusion of this group from most research into pediatric pain has serious implications, as they are particularly susceptible to pain due to their physical disabilities or medical problems associated with them (Nordin & Gillberg, 1996; Ehde et al., 2003; Konstantareas & Homatidis, 1987), to treatments for those problems (Hadden & von Baeyer, 2002), or due to reduced detection or management of typical health problems (Allison & Lawrence, 2004; Hennequin, Faulks, & Roux, 2000). There may also be greater risk for accidental injury in this group (Leland, Garrard, & Smith, 1994), especially those who are mobile (Breau, Camfield, McGrath, & Finley, 2004a). Their injuries may also be more severe and result in more long-term care than those of typical children (Braden, Swanson, & Di Scala, 2003).

Children with cognitive impairments may also have pain for a longer period due to difficulties in detecting their pain and, in some groups, this can have fatal effects (Jancar & Speller, 1994). The complexity of their multiple medical conditions may also make it more difficult to manage pain well, even when it is detected. In summary, there is mounting evidence that pain presents a greater problem for children with cognitive impairments than for typical children, and this is occurring within a growing population, as advancements in medical technology that have increased survival rates and lifespan (Lorenz, Wooliever, Jetton, & Paneth, 1998).

A study aimed at documenting the nature and incidence of the children's pain suggests the pain experienced by this group is not trivial (Breau, Camfield, McGrath, & Finley, 2003b). It reports that pain due to minor accidental injuries had an average rating of 4 on a 0 to 10 scale of pain intensity completed by caregivers, while more common types of pain, due to chronic conditions such as gastrointestinal disorders, musculoskeletal problems, or illnesses, was rated on average at 6.1 out of 10. Given that a score of 30 out of 100 is considered clinically significant (equivalent to 3 out of 10 in this study), and a level at which many parents give medication (Finley, McGrath, Forward, McNeill, & Fitzgerald, 1996), it is obvious that children with cognitive impairments are frequently suffering from clinically significant pain.

Despite increasing evidence that pain is a significant problem for this group, and the emergence of measures to assess their pain, there remains controversy over whether their pain experience differs from that of typical children, with some suggesting problems in pain assessment are due to the fact that individuals with cognitive impairments do not display pain behaviour because they are insensitive to pain (pain insensitivity) or may not interpret what they

feel as negative (pain indifference) (Biersdorff, 1991) (Biersdorff, 1994) (Lu, 1981). However, more recent studies using validated measures suggest that children with cognitive impairments may show more pain behavior (Nader, Oberlander, Chambers, & Craig, 2004). In addition, adults with cognitive impairments may have more difficulty with localizing pain specifically (Hennequin, Morin, & Feine, 2000) and may respond to pain more slowly (Defrin, Pick, Peretz, & Carmeli, 2004), but their sensitivity to pain (Defrin et al., 2004) and reaction to pain may be greater (Porter et al., 1996).

PAIN ASSESSMENT

Against this backdrop of difficulties with pain detection and management, and questions about the actual experience of pain in individuals with cognitive impairments, a small literature has developed that has slowly laid the groundwork for better pain assessment in this vulnerable group of children. The following sections describe the emergence of this literature and summarize what we know about pain assessment in children with children with developmental disabilities and associated cognitive impairments at this time.

First Reports of Observable Pain Response

In 1965, Reynell published the first study that documented the pain response of children with cognitive impairments as part of a study following children with cerebral palsy after surgery (Reynell, 1965). No relation was found between children's pain related behavior and their level of intellectual functioning. This was the first evidence that children with multiple disabilities do display a pain response that can be observed and quantified. A series of papers describing case reports followed over the next two decades. (Mette & Abittan, 1988) (Collignon, Porsmoguer, Behar, Combe, & Perrin, 1992; Collignon, Giusiano, Porsmoguer, Jimeno, & Combe, 1995). Most often they described distinct behaviors that led professionals to investigate the possibility of pain.

Finding Common Pain Behaviors

Only in 1995, 30 years after Reynell's original study (Reynell, 1965), did the next study appear that attempted to quantify observable pain behavior in individuals with cognitive impairment (Giusiano, Jimeno, Collignon, & Chau, 1995). This group developed a list of behaviors based on observations during a physical exam of 100 residents, aged 2 to 33 years, residing in a long-term care facility. All were nonverbal and 70% were described as having a "chronic vegetative state". This ground-breaking study provided the first evidence of common pain behaviors in this population. One difficulty with the results, however, was that the items generated were based on change from "usual" behavior, making the tool difficult to use for people not familiar with an individual.

The same year, Hunt and Burne reported on parents' perceptions of the pain behaviors of 120 people aged 1 to 25 years with static or progressive encephalopathies (Hunt & Burne, 1995). The most common signs of pain included postural and physiological changes, crying, and facial expression. The majority of parents also reported their child's mood changed with pain.

The following year, Fanurik, Koh, Harrison and Conrad reported a study of 66 children with cognitive impairments (Fanurik, Koh, Harrison, & Conrad, 1996). In this case, 41% of children had severe cognitive impairments, but 48% did have verbal skills. Parents' descriptions of their child's reaction to a previous needle stick included: verbalization, localization of the painful area, cry, behavioral or emotional changes, facial expression, body movement and self-abusive behavior. Unfortunately, the retrospective nature of the study limits conclusions.

This group later interviewed a set of parents regarding their children's pain experience, expression and treatment (Fanurik, Koh, Schmitz, Harrison, & Conrad, 1999a). Parents' descriptions of how their child displays pain varied with the level of their child's cognitive impairment. For example, 57% of parents of children with mild or moderate impairments said

their child made direct verbal statements about pain, but only 7% of parents of children with severe to profound cognitive impairments reported this. Many parents also felt their child's pain was underestimated or under-treated by healthcare professionals (Fanurik et al., 1999a). Twenty-nine percent believed their child's pain was treated differently because of their cognitive impairments.

Investigating Physiological Pain Response

Another avenue investigated has been the possibility that there may be physiological responses to pain that could be used to detect pain, especially when children with cognitive impairments have very limited, or no verbal ability. In one study, response to pain in eight adolescents with spastic quadriplegia (Oberlander, Gilbert, Chambers, O'Donnell, & Craig, 1999) was evaluated as they received both a mock and a real injection in random order. No significant increase in heart rate was found during the injection. Nor did facial action increase. Although these results suggest physiological measures may not be sensitive to pain for children with altered neurological systems, the sample size was small, making it possible that there was insufficient statistical power.

Research with adults with similar impairments also suggests Oberlander et al.'s results may not reflect pain insensitivity (Oberlander et al., 1999), but rather, a lack of physiological reactivity to pain. One recent study found relatively lower heart rate and blood pressure response to a cold pressor test, as well as to a hand-grip test, in individuals with cognitive impairments than in typical controls (Fernhall & Otterstetter, 2003). The authors suggest this reflects a general decreased sympathetic modulation in this population. A similar attenuation of autonomic response to pain was found by another group when they investigated the pain response of adults with Alzheimers (Rainero, Vighetti, Bergamasco, Pinessi, & Benedetti, 2000). In a subsequent study, they found that pain sensation was unrelated to brain electrical activity or cognitive status (Benedetti et al., 2004). In contrast, they found that heart rate increase to pain was related to delta and theta frequencies. They conclude that pain sensation is not altered by cognitive status, while autonomic status is. Porter et al. report a similar blunting of heart rate increase in response to venipuncture in a sample with Alzheimers (Porter et al., 1996). Again, in their sample, behavioral response to pain was not diminished.

In summary, the bulk of research to date suggests that physiological response to pain in individuals with cognitive impairments may be reduced or qualitatively different relative to other groups. However, this appears to reflect differences in the mounting of response to pain, rather than a decrease in pain sensation. This idea that there may be a disconnect between pain sensation and pain response in some individuals with cognitive impairments is supported by a very recent study of adults. In that study threshold for heat pain was increased relative to controls if a measure that depended on response time was used. In contrast, when a measure of threshold that was independent of response time was used, those with cognitive impairments displayed reduced conduction velocity and reaction time, but lower pain thresholds (Defrin et al., 2004).

Thus, at this time, physiological responses to pain do not appear promising as a method of clinical pain assessment. It appears those with cognitive impairments may have altered responses to many external stimuli, and that this reflects differences in physiological response, but not pain perception. Given the heterogeneity of this population, it may be some time before we have an indication of whether reliable physiological responses can be used to detect or measure pain.

Research of Pain Assessment Tools Designed for Typical Children

Measures of Facial Reaction to Pain

Another approach to pain assessment in children with cognitive impairments that has been explored is facial expression. Research suggests typical children display a facial response to pain (Breau et al., 2001). Oberlander et al. were the first to explore this possibility, in their study of adolescents described previously (Oberlander et al., 1999). However they found no increase in

facial activity in response to pain using the Child Facial Coding System (Chambers, Cassidy, McGrath, Gilbert, & Craig, 1996). This may reflect their methodology. They chose to analyze only facial action occurrence, through summing the occurrence of all actions over each period of time examined. Previous research suggests the intensity of facial action carries important information regarding pain, and inclusion of this parameter may have altered the results (Breau et al., 2001).

Other studies have found facial responses to pain in groups with cognitive impairments. Porter et al. report the elderly adults in their study with Alzheimers displayed a greater, but less easily classified, facial response to pain than those without cognitive impairments (Porter et al., 1996). Similarly Nader et al. found increased facial activity in children with autism relative to controls, when they were observed during venipuncture (Nader et al., 2004). Mercer and Glenn also studied facial response in infants with and without developmental delays (Mercer & Glenn, 2004). They found facial response was not diminished, but was more diffuse in the infants with impairments.

Overall, this small literature suggests facial activity may have merit as a method of pain assessment for children with cognitive impairments. However, there is clearly a great deal of work to be done in this area before clinical use is feasible. Those with impairments may not show the same pattern of facial response as typical children, necessitating the development of new coding schemes for deciding which pattern does reflect pain. Most current systems are also time-consuming and coded from film, making bedside application impractical at this time.

Multidimensional Pain Assessment Tools

Several studies have examined the validity of multidimensional pain tools designed for typical children when used with those who have cognitive impairments. Voepel-lewis et al. investigated the validity of nurses' scores for 79 children on the Face, Legs, Activity, Cry, Consolability (FLACC) scale for postoperative pain in children with cognitive impairments (Voepel-Lewis, Merkel, Tait, Trzcinka, & Malviya, 2002). Parents also provided estimates of their child's pain using a visual analogue scale (VAS) of pain ranging from 0 to 10. Correlations between FLACC scores and parent VAS ratings ranged from .52 to .65, although parent ratings appeared to be higher. FLACC scores decreased significantly with administration. However, children with mild pain who were not administered analgesics were omitted from these analyses. Thus, caution must be taken in interpreting this finding as it can only speak to reductions in scores for children with moderate to severe pain. Likewise, the authors report that agreement between observers was not as good for moderate pain as for mild or severe pain. This suggests observers were able to use the scale to distinguish very low versus very high pain, but may have had more difficulty when pain was at neither extreme.

Soetenga et al. used the University of Wisconsin Children's Hospital Pain Scale and the Wong-Baker Faces Scale (Wong & Baker, 1988) with 74 children admitted to hospital, 15 of whom were over age 3, but nonverbal (Soetenga, Pellino, & Frank, 1999). This scale contains four subscales: Vocal, Facial, Behavior and Body Movement. The correlation between the two was .53 for parents and .89 for nurses. Scores were significantly lower after analgesic administration and inter-rater reliability was excellent. Unfortunately, the authors do not report subgroup analyses for the nonverbal children in the sample, so it is difficult to conclude whether the psychometrics presented would be similar had the scores for the subgroup been examined separately.

Most recently, Solodiuk and Curley suggest the use of an individualized visual analogue scale (VAS) for children with severe cognitive impairments (Solodiuk & Curley, 2003). A caregiver is asked to provide descriptors for the 0 and 10 anchors and points between. They report this has worked well clinically, but have not yet reported data on the validity or reliability of this adaptation to a tool commonly used by typical children.

Overall, there is not convincing evidence that the tools above are satisfactory for children with cognitive impairments. Although further research may provide additional evidence to support their use clinically, they are not recommended at this time.

The Emergence of Multidimensional Pain Tools Specifically for Children with Cognitive Impairments

Since Giusiano et al.'s first attempt to determine a common set of pain behaviors in institutionalized individuals with severe cognitive impairments (Giusiano et al., 1995), several groups have developed multidimensional pain assessment tools specifically for children with cognitive impairments. Although there is some overlap between these, each also has its unique characteristics, and some have focused on broader populations (e.g. children and adults), while others have primarily looked at a specific subgroup (e.g. institutionalized individuals) or situation (e.g. postoperative). Table 1 depicts the items contained in these, which gives a sense of the similarities.

Echelle Douleur Enfant San Salvadour (DESS)

Since their first study published in English in 1995, Collignon and Giusiano and their group have continued their development of the DESS. In a second study published in English, they report refinement of the scale (Collignon & Giusiano, 2001). After reducing the number of items to 10, they report that a score of 2 indicates attention should be paid to the child, as pain is possible. A score of 6 on their scale, they report, indicates definite pain that requires treatment. Two problems exist with the items of this scale. First, there is overlap between items. For example, the addition of crying or jerking to some behaviors (e.g. protection of painful areas) increases scores for that item. Since crying and jerking are also contained in specific items, this means scores for the items are not independent, undermining assumptions for statistical analyses. Second, an item that involves the individual seeking a comfortable position also includes the possibility that the nurse placed the person in that position. Thus, the judgment of the nurse becomes incorporated in behavioral ratings. Unfortunately, the DESS is also designed so that items are rated in relation to the individual's typical behavior. This may be possible, or even preferred in situations where an adult familiar with the individual is assessing pain, such as in the home setting or large, long-term residential centers. However, this format is not appropriate in other situations, such as in emergency situations or in hospital where staff may be frequently change. Using change from typical behavior also means that scores cannot be used across situations (e.g. home versus school/hospital) as children's usual behavior may differ across these settings. Finally, validation of the DESS was conducted in French. Caution should be taken in using those cut-off values if using an English version of the scale until systematic translation and validation in English is completed, as cultural and language differences can impact both pain behavior and our perception of pain behavior.

The Non-Communicating Children's Pain Checklist

McGrath and his group initiated a program to develop an observational pain tool specifically for children with severe impairments in the mid-1990's (Breau, McGrath, Camfield, Rosmus, & Finley, 2000). Items were generated through semi-structured interviews with the primary caregivers of 20 individuals aged 6 to 29 years (McGrath, Rosmus, Camfield, Campbell, & Hennigar, 1998). Thirty-one behaviors were extracted and grouped into seven subscales (Vocal, Eating/Sleeping, Social/Personality, Facial Expression, Activity, Body & Limbs, and Physiological). Although specific behaviors varied amongst individuals, all individuals displayed some behavior from each subscale.

In a subsequent study, the new pain measure, the Non-communicating Children's Pain Checklist was validated in a home setting (Breau et al., 2000). One item was dropped, due to low endorsement, leaving 30 items that were rated as present or absent. Thirty-three caregivers indicated whether each item was present for four observations, two in which pain was present,

one in which the individual with cognitive impairments was distressed but had no pain, and one calm period. An important aspect of this study was that the pain events observed by the caregivers were heterogeneous, including both acute pain, such as bee stings, minor burns, falls and intravenous insertions, and longer-term pain such as severe burns, throat/urinary tract/sinus infections and postoperative pain. The results indicated scores were consistent over the two pain events, despite the causes of pain differing in most cases. Scores during calm were also significantly lower, suggesting the tool was sensitive to pain.

In a second study, information was collected from caregivers about their children's typical pain behavior in order to predict behavior during a subsequent pain event (Breau, Camfield, McGrath, Rosmus, & Finley, 2001). A core set of NCCPC items that were reported as "typical" by caregivers had significant odds of appearing during subsequent pain episodes. The seven items were: "not co-operating, cranky, irritable, unhappy", "seeking comfort", "change in eyes", "less active", "gestures to part that hurts", "tears", and "sharp intake of breath". A multiple R of .70 indicated the seven items also played a significant role in predicting pain intensity, while a nonsignificant R of .31 in predicting distress, suggested the items were specific to pain. A replication, in which 63 new caregivers' ratings of these seven items were used to predict the presence of pain, provided additional evidence that these behaviors are particularly consistent. The presence of the seven NCCPC items during the subsequent episode correctly identified the presence of pain in 69% of cases and the absence in 95% of cases.

A third study of the NCCPC investigated its use for postoperative pain (Breau, Finley, McGrath, & Camfield, 2002). Items from the Eating/Sleeping subscale were excluded and more detailed responses from observers were added, such that they now provided ratings to indicate whether each item was observed "not at all", just a little", "fairly often" or "very often" during 10 minute pre- and postoperative observations of 24 children. Caregivers' and researcher' total scores were significantly higher after surgery. The correlation between caregiver and researcher postoperative NCCPC-PV scores was .72, indicating good inter-rater reliability. A score of 11 was found to be best for detecting moderate to severe pain. The equivalent 100 mm VAS ratings for children with mild pain were 12.0 ($SD = 11$) and for moderate to severe pain, 53.5 ($SD = 17$).

The new NCCPC was also re-examined in a home setting with a larger sample than the early studies (Breau et al., 2000). This 30-item version also incorporated the rating system developed for the postoperative version (not at all, just a little, fairly often, very often). Seventy-one caregivers completed the Non-communicating Children's Pain Checklist-Revised (NCCPC-R) for two-hour observations of their child at home. Scores for children who experienced pain during the observations were compared to scores for children who did not have pain. Pain in this group was due to a wide variety of causes, including falls, self-injury, gastrointestinal reflux, constipation and surgery. Fifty-two caregivers also completed the NCCPC-R for a second episode of pain to assess consistency in scores across pain episodes. Scores during pain for the two pain episodes differed significantly from scores when pain was absent. A total score of 7 had very good sensitivity (84%) and good specificity (65%- 77%) for detecting any pain. The results also indicated total NCCPC-R scores were consistent across the two episodes of pain, as were the number of items children displayed. The number of items displayed were consistent with those reported in the first study (Breau et al., 2000), indicating good consistency across samples. Because of concerns that children display pain erratically, further analyses of children's individual consistency in scores was conducted. These indicated 93% of the 55 children who were observed during two episodes of pain had scores during their second observed episode of pain that fell within 95% confidence intervals of the scores they received for their first episode of pain.

The NCCPC's were specifically designed for children who had very limited verbal abilities due to their cognitive impairments. However, Hadden and von Baeyer have also used the NCCPC-R with children with cerebral palsy with varying levels of verbal abilities. Parents identified items from the checklist that occurred when their child had pain. The reported presence

of 24 items from the NCCPC-R did not vary due to the children's communication ability (Hadden et al., 2002). However, caregivers of children who could not communicate verbally reported a greater frequency of several items, including "decreased sleep", "jumping around, agitated, fidgety", "stiff, spastic, tense, rigid" and "lips pucker up tight, pout, quiver". On the other hand, caregivers of children who could communicate verbally reported a greater frequency of the items "gestures to or touches part of the body that hurts" and "protects or favors part of body that hurts". Thus, it appears the children displayed different patterns of items, but achieved similar total scores when parents use the tool retrospectively.

In a subsequent study, Hadden and Von Baeyer investigated the scores of 129 children with cerebral palsy, with a wide range of verbal abilities, during home physiotherapy exercises that were expected to cause pain (Hadden & von Baeyer, 2001). Although children's facial expression did not change significantly during active stretching, scores on the NCCPC-PV were significantly higher. Thus, there is some evidence that the NCCPC's may be valid for higher functioning children with physical impairments. However, further research is needed to examine possible differences in sensitivity and specificity for higher functioning groups before clinical use with these groups is recommended.

Breau et al. also examined whether the NCCPC-R is valid for children who display self-injurious behavior (Breau et al., 2003a). This is a concern because there is a greater belief that these children may be insensitive to pain (Gillberg, Terenius, & Lonnerholm, 1985; Sandman, Barron, Chicz-DeMet, & DeMet, 1990) or display pain differently. There were no significant differences in NCCPC-R total scores for observed episodes of pain in 101 children with ($n = 44$) and without ($n = 57$) self-injury. In fact, children who self-injured received nonsignificantly higher total NCCPC-R scores and higher scores on the Vocal and Social subscales. The results also suggested that children who had chronic pain might show a different pattern of self-injury, suggesting self-injurious behavior might be a reaction to pain in some children, rather than evidence of insensitivity to pain.

Nader et al. also used the NCCPC-R in their study of venepuncture pain in children with autism (Nader et al., 2004). In this case, parents were asked to complete the NCCPC-R retrospectively for a past pain event. Unexpectedly, scores for the past event were negatively correlated with facial response to pain during the venepuncture, as well as ratings of distress using the Observational Scale of Behavioral Distress (Jay, Ozolins, Elliott, & Caldwell, 1983). The authors suggest this finding may reflect the inappropriateness of the two measures of venepuncture pain, or the fact that children with autism may display very different behaviors in strange or new situations. Given the mixed results described previously regarding the facial display of pain in several samples with cognitive impairments, the former explanation may be valid. However, it is also possible that the children reacted atypically during this event, especially as they were restrained through bundling with a blanket by adults they were not familiar with. This may have resulted in a great deal of distress, distracting them from the actual pain due to venepuncture. Thus, it may be that the behaviors observed were more specific to distress than pain.

At this time, eight studies have included one of the NCCPC's, with five specifically examining the psychometrics of the two tools for everyday pain and for postoperative or acute pain (NCCPC-Revised, NCCPC-PV, respectively). The data supporting the validity and reliability of the tools looks promising, and cut-off scores have been developed, making the two useful clinically. One problem with the validation of the NCCPC-R is that it was conducted with 2-hour observations, impractical in a hospital setting. We have recommended that caregivers use the NCCPC-PV and 10-minute observations if observations of 2-hours are not feasible. As with all pain assessment tools, however, repeated evidence is required with different samples and in different situations to provide cumulative support for clinical use. Currently, evaluations of several translations of the NCCPC's are underway (French, Swedish, German) and these will provide additional information regarding the situations and populations for which the NCCPC-R

and NCCPC-PV are valid. Research examining the validity of the NCCPC's with subgroups of children, such as those with Batten Disease, are also planned.

The Pain Indicator for Communicatively Impaired Children (PICIC)

Stallard et al. developed the Pain Indicator for Communicatively Impaired Children (Stallard et al., 2002). Numerical (0-5) pain ratings of 49 caregivers for their children were predicted using ratings parents provided of six different pain cues, rated as occurring "not at all", "a little", "often" or "all the time", as well as individual pain cues identified by caregivers. Five of the six cues proposed by Stallard's group achieved 67% sensitivity, three of the cues were associated with ratings of pain severity on a 1-5 scale, and twenty additional pain cues were identified by caregivers.

Unfortunately, cut-off scores have not yet been developed for this scale and inter-rater reliability was not assessed in the study. The authors also do not provide information about the time frame of the observations upon which the parents based their ratings. However, the scale is short, making it quick to use if further studies provide additional information regarding its psychometric properties.

The Paediatric Pain Profile

The Paediatric Pain Profile was developed through interviews with 21 caregivers of children with cognitive impairments (Hunt, Mastroyannopoulou, Goldman, & Seers, 2003). From an original pool of 56 items, a set of 20 was selected for the final version. Items are rated as occurring "not at all", "a little", "quite a lot" or "a great deal". The scale was examined in a subsequent study (Hunt et al., 2004). When asked retrospectively, parents' ratings of their child on the scale were significantly higher with respect to a typical pain the child had than when the child was "at their best", and they correlated with parents' Visual Analogue Scale pain ratings for that typical pain. Simultaneous ratings by 54 parents and another observer familiar with the child in the home indicated inter-rater reliability was good (.73). Forty-one parents also provided ratings for their child prior to and after administration of a short-acting analgesic in the home. Scores were significantly lower after analgesics were given. Finally, scores were examined in 30 children having gastrointestinal or orthopedic surgery. However, scores did not decline over from the first to the fifth postoperative day.

This set of studies provides some preliminary evidence of the validity and reliability of the Paediatric Pain Profile. Although the inter-rater reliability appears good, it was based on ratings from two adults familiar with the child. Further research is needed to determine if the Paediatric Pain Inventory displays similar reliability and validity when used by professionals who do not know the children being rated well. Similarly, although a cut-off score is provided, it was developed through averaging the pain ratings of two observers familiar with the child, something that would not be available in most clinical settings. Further, 96 pairs of ratings were used to develop the cut-off scores, but it appears only 54 of these were conducted simultaneously and the time frame of the observations is unclear. The scale should be simple to complete, making clinical use feasible. However, the unclear time frame for observations is a problem in terms of determining the length of time upon which observations should be based. In addition, several items are reversed scored, adding somewhat to the task of computing scores. One positive attribute of the scale is that the developers have generated a package that caregivers can complete, documenting their child's baseline behaviour and behaviour during common pains. This might be particularly helpful in cases where children have multiple conditions that cause chronic or recurrent pain.

Other Scales in Development

While the Pain Indicator for Communicatively Impaired Children and the Pediatric Pain Profile have focused primarily on pain outside the hospital setting, another group has developed a

pain assessment tool for postoperative pain. Terstegen and her colleagues generated 209 potential items through interviews with parents and professionals who work with children with cognitive impairments, through observations of children during events expected to be painful, such as physiotherapy, vaccinations and dental treatment, and through review of the literature (Terstegen, Koot, de Boer, & Tibboel, 2003). This pool of items was reduced to 23 that appeared sensitive to pain in 52 children who had surgery. They also found 13 items achieved higher ratings during pain due to a procedure than after surgery, suggesting a possible difference in pain display depending on whether pain is short / acute, or longer lasting. Few additional psychometrics of the scale are presented. Thus, this tool is not yet appropriate for clinical use. However, it shows promise for differentiating pain types.

In all, five tools have been developed specifically for children with cognitive impairments in the space of only a few years. Each has been developed by a different group, and through slightly different methods. Interestingly, there is a great deal of overlap in the items included in each (Table 1). For example, all items of the Pain Indicator for Communicatively Impaired Children (Stallard et al., 2002) are included in the NCCPC's (Breau, McGrath, Camfield, & Finley, 2002; Breau et al., 2002), the Paediatric Pain Profile (Hunt et al., 2004), and the tool in development by Terstegen et al. (Terstegen et al., 2003). Twelve items from the NCCPC's are also included in Terstegen et al.'s new tool, and all but 2 of the 20 items of the Paediatric Pain Profile assess behaviours included in the NCCPC's. It is also notable that three of the four tools incorporate between 20 and 30 items. This supports the notion that children with cognitive impairments, especially those with severe impairments and limited verbal ability, may exhibit a core group of pain signs across situations.

At this time, the NCCPC-Revised and NCCPC-Postoperative Version have accumulated the most support for their psychometric properties and these scales are used clinically to supplement clinical judgment. Several large scale studies of English, French and Swedish versions of the tools are also underway, aimed at providing new information about their use and possibly refinements to make them more feasible in clinical practice. For example, in several studies, assessments are being made using observations of only five minutes to determine if reducing the observation period will affect their sensitivity to pain. It is also hoped that more cut-off scores can be developed with these larger groups so that scores can be equated with points on a Visual Analogue Scale of pain intensity.

That fact that so many groups have begun development of scales in such a short period of time highlights a growing recognition of the difficulties clinicians and parents encounter in judging pain in children with cognitive impairments. Almost all the children involved in these studies had moderate to severe cognitive impairments, with little or no ability to communicate their pain verbally. However, many children have milder impairments and may be able to provide some indication of their pain. The literature investigating the reliability of their self-report, however, consists of only a few studies.

Self-Report of Pain

Fanurik et al. were the first group to investigate the self-report abilities of children with cognitive impairments (Fanurik, Koh, Harrison, Conrad, & Tomerlin, 1998). Their study included 47 children with varying levels of cognitive impairments and 111 children without impairments scheduled for surgery. Children's ability to use a 0 – 5 numerical rating scale of pain was assessed through several tasks. Children's ability to understand the concepts of magnitude and order were tested using tasks in which they were required to order wooden blocks by size and to arrange numerals in order. Children were then asked to match cards depicting faces at different levels of pain with cards depicting pain intensity (1, 3 and 5).

Only 10 (21%) of the children with cognitive impairments could complete all three tasks, and all had mild to borderline impairments. An additional 23 (44%) of the children could complete some part of the tasks. None of these completed the final task of assigning numerals to

faces depicting pain intensity. In contrast, all children without impairments who were above age 8 completed all tasks, while 18% of children without impairments who were between age 4 and 7 were able to complete all tests and 32% completed at least some tasks.

An interesting secondary analysis in this study examined nurses' ability to predict which children were capable of using a numerical rating scale. Only 47% of children with cognitive impairments who were deemed capable by nurses did demonstrate the ability. In most cases, nurses tended to overestimate the abilities of children with mild to moderate impairment. Nurses also overestimated the skills of 33% of children without impairments.

More recently, Benini et al. describe a study of the self-report skills of 16 children aged 7 to 18 years with mild to moderate cognitive impairments (Benini et al., 2004). In this novel study, children were administered a one-hour training on use of the pain tools used prior to receiving a venepuncture. Children then completed original and adapted versions of a 10 centimeter Visual Analogue Scale of pain, the Eland Color Scale (Eland, 1985), which depicts a picture of a body that the child uses to indicate pain location, and the Faces Scale (Wong et al., 1988). There were no differences in children's ability to use the scales based on level of impairment (mild, moderate) or diagnosis (tetraplegic, Down's Syndrome). The authors report that more children completed the modified scales (numbers of faces in Faces Scale reduced; body parts enlarged on Eland Color Scale; set of 5 cubes replaces 10 cm Visual Analogue Scale). Unfortunately, relations between parent ratings of pain and each instrument were not provided, but there was a moderate consistency between the Visual Analogue Scale ratings provided by parents and a researcher and those provided by children.

Only one other study has examined the self-report skills of children with cognitive impairments. Zabalia et al. (2005) investigated the ability of 14 children with mild to moderate cognitive impairments aged 8 to 18 years to use a 100 mm Visual Analogue Scale of Pain and a the Faces pain Scale-Revised (Hicks, von Baeyer, Spafford, van, I, & Goodenough, 2001) to rate the pain of vignettes and the pain they believed they would experience were they to experience the event in the vignettes. The vignettes depicted individuals in pain due to a burn from a casserole, falling from a bicycle, falling from roller skates and a vaccination. After ratings were provided, the children were also asked to describe the quality of the pain each event would cause.

The children's pain ratings for both the vignettes and the pain they would feel in that situation did vary by the cause of pain, suggesting they did distinguish between the pain events. They also provided up to nine words to describe the quality of the pain for the events, a number that is similar to that reported by typical children of a similar mental age, and the words were appropriate for the pain depicted (e.g. burning, pinching). One weakness of the study is that the children did not rate their own pain for an actual experienced event. However, the results do indicate the children had some skill in rating pain, and most importantly, that they could describe pain quality.

In summary, there is only a small body of research evaluating the ability of children with cognitive impairments to provide self-report of pain. This indicates that children with mild impairments may be able to use some self-report tools to provide a first-hand rating of the intensity of their pain. The research also suggests children with mild to moderate impairments can distinguish between different types of pain and may be able to provide descriptions of the quality of their pain that could be useful for diagnosing pain cause. Clinicians should investigate each child's abilities to use self-report tools, as it appears they may overestimate children's skills. Using simple analogue tasks may be feasible in some situations. In addition, clinicians should request estimates from parents or caregivers in addition to child self-report, or use observational pain tools, until further evidence is available to show that children can provide reliable self-report in clinical situations that may be more distressing (and, possibly, more painful), or until the psychometric properties of modified tools, such as those described by Benini (Benini et al., 2004), have been evaluated more fully.

The Role of the Observer in Pain Assessment

Parents have identified pain as one area of concern for their children who cannot communicate verbally (Stephenson & Dowrick, 2000). They also express concern that some professionals may discount their reports that their child has pain (Hunt et al., 2003) and that their child's pain is treated differently than that of typical children (Fanurik et al., 1999a).

In interview studies, parents also report that familiarity with their child is required to assess the child's pain (Carter, McArthur, & Cunliffe, 2002; Hunt et al., 2003). No scientist or clinician would argue the fact that each child is unique. However, parents may be basing this assumption on past experience in which professionals failed in attempts to discern pain due to lack of training, lack of literature to provide guidance, and lack of structured tools to use. Our knowledge of the process of developing structured tools, and of gathering evidence for their validity has, however, grown. And, variation in individuals' pain behavior does not preclude the use of measures that have been empirically and scientifically tested for clinical use. We must keep sight of our primary goal, which is not to generate a precise picture of the child's pain experience, but to have sufficiently accurate information upon which to base treatment decisions. For most professionals, a tool that can distinguish between no pain and mild, moderate or severe pain is sufficient for their everyday practice.

Research also suggests that, when provided with information in a structured way, or using a validated tool, observers who are unfamiliar with children with cognitive impairments may provide very reliable judgments. For example the good inter-rater reliability between parents and a researcher reported by Breau et al. (Breau et al., 2002) suggests use of a structured tool may facilitate agreement between parents and an unfamiliar observer. Two recent studies by Stevens' group also suggest that structured information and tools may also help to minimize pre-existing beliefs that observers may have that could impact judgments of pain in those with cognitive impairments. In one study, professionals who completed a questionnaire asking about the pain of infants at risk for cognitive impairments indicated a belief that greater impairment lead to a reduced pain experience (Breau et al., 2005). In contrast, when presented with the task of assessing the pain of infants at varying levels of risk for neurological impairment from videotape, using 0-10 Visual Analogue Scale pain ratings, the pain ratings of a similar group of professionals did not vary due to descriptions of the infants' level of risk for neurological impairment (Breau et al., 2004b). Similarly, when presented with vignettes, pain ratings by professionals in a study by Fanurik et al. also did not differ due to the described level of cognitive impairment of the children (Fanurik et al., 1999b).

To date, we do not know if biases or pre-existing beliefs affect adults' judgments of pain in children with cognitive impairments. A belief that individuals with cognitive impairments have a reduced pain experience has been reported for caregivers (Breau, MacLaren, McGrath, Camfield, & Finley, 2003) and professionals (Breau et al., 2004b). However, it is not clear whether these operate through effects on pain assessment or management, or whether professionals are able to set these aside in clinical situations. This is an important area for future research if optimal care is to be provided for this very vulnerable group.

Clearly, assessment is only the first step in the process of alleviating a child's pain. While the literature regarding pain assessment for this vulnerable group is small, that regarding pain treatment is almost nonexistent. The next section of this chapter describes issues regarding treatment of acute and chronic pain in children with cognitive impairments, and highlights the difficulties clinicians face in their efforts to manage what can be very complex pain problems.

TREATMENT OF PAIN

There has been very little research on treatment of pain in infants or children with cognitive impairment. Almost all studies of treatments of pain have excluded these children. This is in spite of the fact that they are more likely to have painful conditions and more likely to have

painful procedures than other children (Breau et al., 2003b; Stallard, Williams, Lenton, & Velleman, 2001).

There are two major strategies for determining what treatment to use for pain in children with cognitive impairment. The first and the preferred method of treatment should be to diagnose the cause of the pain and treat this underlying cause. The second approach is to symptomatically treat the pain without knowing its cause.

Acute Pain

For procedure pain and for postoperative pain, the diagnosis is usually obvious. Lacking any evidence, we should assume that, in general, treatments that are effective in typical children will be effective in children with cognitive impairments. However, a recent study of infants at risk for neurological impairment in a NICU setting suggests this may not always be the case. In their study of 194 infants, Stevens et al. report that infants with greater risk for neurological impairment received fewer analgesics for procedure-related pain on the first day of life (Stevens et al., 2003). They also found that analgesic administration was related to the number of procedures performed for most infants, but not those at highest risk for future impairments.

Many treatments found effective for typical children could easily be implemented with children with cognitive impairments. For example, the use of EMLA®, an eutectic mixture of lidocaine and prilocaine is likely to be effective in reducing pain from needles in children (Halperin, McGrath, Smith, & Houston, 2000). Similarly, distraction is effective in reducing pain from short procedures (Cohen, Blount, & Panopoulos, 1997).

However, treatment may not always be straightforward. There may be more problems in implementing treatment in the child with cognitive impairments than in other children. In the case of EMLA® for example, some children with cognitive impairment will not tolerate the EMLA® patch for an hour before the needle. Some children with cognitive impairments may be so distressed by the medical situation that they are difficult to distract. Adults who are unfamiliar with a child may also find it difficult to determine that child's developmental level, in order to decide upon an activity that could be distracting. Modeling, shaping, and other behavioral techniques may be helpful in reducing children's distress and optimizing cooperation. Souder et al. provide an excellent description of techniques used for children with autism spectrum disorders (Souders, Freeman, DePaul, & Levy, 2002), based on experiences during a large trial which entailed venepuncture (Levy et al., 2003). Many of these may be effective with other children with cognitive impairments.

Post-operative pain management for children with limited cognitive abilities, as well, generally follows the strategies that are useful in other children with a few exceptions. The selection, dosages and schedules of drugs used should be no different from other children. Specific challenges may arise. Children with airway problems may need to be monitored even more carefully than a child without any difficulties. Several case studies have reported specific instances of anesthesia management in children with particular syndromes which involve cognitive impairments (Shenkman, Krichevski, Elpeleg, Joseph, & Kadari, 1997; Adhami & Cancio-Babu, 2003; Iacobucci, Galeone, & De Francisci, 2003; Courreges, Nieuviarts, & Lecoutre, 2003). (Critchley, Gin, & Stuart, 1995). Children with cognitive impairment are unlikely to be able to use patient controlled analgesia. However, successful use has been reported for typical children over five years of age (McDonald & Cooper, 2001; Birmingham et al., 2003). Thus, with older children with cognitive impairments with mental abilities greater than a typical five-year-old, patient controlled analgesia may be possible.

There is evidence that children with cognitive impairments do not receive the same care as children without cognitive impairments postoperatively. In one of the few studies of postoperative pain management in children with cognitive impairment, Malviya et al. (2001) compared the pain assessment and management practices in 42 children (19 with cognitive

impairment and 23 without impairment undergoing spinal fusion (Malviya et al., 2001). They found that children with cognitive impairment received smaller total opioid doses.

More recently, Koh et al., (2004) prospectively examined 152 children with cognitive impairment of different severities and 138 non impaired children . They found that children with cognitive impairment undergoing surgery received less opioid in the perioperative period than children without cognitive impairment. Children with cognitive impairment were given similar amounts and types of analgesics in the postoperative period as children without cognitive impairment. Koh et al. (2004) suggested that anesthesiologists believe that children with cognitive impairment are more sensitive to the side effects of opioids than other children. Unfortunately, there is no research investigating this phenomenon.

In summary, there is a dearth of data on acute pain in infants and children with cognitive impairment that can guide their care and they may receive fewer analgesics than children without cognitive impairment. The use of nonpharmacological treatments has been most neglected, and, yet, might offer many options for children's acute pain when their complex conditions raise concerns about analgesic administration.

Chronic and recurrent pain

Diagnosis of the Etiology of Pain

Clinicians are well aware of the methods of diagnosis and diagnosis in relationship to pain in children with cognitive impairment is fundamentally no different than diagnosis of other children. However, it is important to emphasize issues that are particularly important with this population. These issues arise because children with severe cognitive impairment cannot verbally communicate and because many of these children often suffer significant co-morbidities, especially speech defects, epilepsy and cerebral palsy (Arvio & Sillanpaa, 2003).

The first step in diagnosis is to take a thorough history of the pain problem and the child. In this population, history must be taken almost exclusively from the adults in the child's world. Continuity of care may be even more important in these children than in other children because knowledge of the child's typical problems and usual behavior may help in diagnosis. On the other hand, each problem that every child presents with deserves a careful and detailed analysis. It is also important to keep in mind the child's abilities and estimated developmental level, as children with cognitive impairments may display pain behavior that more closely resembles mental age peers than chronological age peers and caregivers may not always be aware that these behaviors may reflect pain.

A problem arises if the accompanying adult is not well acquainted with the history of the child's current problem. This may be the case if the child lives in an institution such as a group home. In these situations it is important to obtain supplementary information from caregivers who know the child by telephone. Encouraging the child's caregivers to keep written reports of any problems will help.

The importance of a thorough physical examination cannot be over emphasized with this group of children. Children must be undressed, taken out of their chairs and thoroughly examined. Because children cannot verbally localize pain for the clinician, it is imperative to examine the entire child. Children with cognitive impairment may be fearful of examination and it is often necessary to proceed gently and slowly with the physical examination so as to avoid causing distress.

Children with significant cognitive impairment are at higher risk for several conditions that do not as frequently present in children with normal cognitive functioning. Clinicians should have an increased index of suspicion for these problems. In a two-round Delphi poll we (Choo et al., Unpublished) surveyed an international panel of clinicians with experience with pain in children with significant cognitive disability (Choo, McGrath, Finley, Camfield, & Breau, 2001). We found good concordance among these clinicians that gastrointestinal problems were a major source of pain. They also agreed that children with cognitive impairment frequently suffer

musculoskeletal pain, pain as the result of infection, headache, pain from skin problems, neuropathic pain, dental pain and pain from self-injury. Other research by our group indicates children's characteristics may also heighten their risk for specific types of pain (Breau et al., 2004a).

Although recommendations for use of a directed history and behavioral changes for diagnosis of particular health problems has been presented in a well-written paper by Bosch (Bosch, 2002), and Tracy has outlined the ways in which developmental delays may impact the presentation of physical conditions (Tracy & Wallace, 2001), there has not been a systematic mapping of the painful conditions that children with cognitive impairment suffer from. This type of mapping would be of considerable value to the clinician. We are currently using the tacit wisdom of expert clinicians who deal with these children to develop clinical algorithms to help in the diagnosis of different causes of pain. This web-based program will work from symptoms to suggest possible diagnoses that may be relevant. Or it will give symptoms for a chosen diagnosis. In addition, the algorithm will indicate predisposing factors and treatment options.

For example, a clinician might indicate that their patient appears to be in pain and has a cough, fatigue, is floppy and irritable. A list of diagnoses that these symptoms could be associated with is produced. In this case, 4 symptoms match pneumonia and 2 symptoms match gastroesophageal reflux, osteomyelitis, septic arthritis and inflammatory arthritis. The clinician can then look at the other symptoms for that diagnosis and narrow down the diagnosis. Different tests are suggested as possibilities.

The algorithm is still under development and testing. It will not diagnose problems but can provide prompts for clinicians who may not be thinking of a possibility.

Treating pain without knowing its cause

Pain clinicians treating adults or children with pain often cannot find the cause of the pain. This is also true with children with cognitive impairment. In these situations, the treatment will be symptomatic. General principles of good clinical care are used. Interventions are chosen on the basis of an analysis of what is known.

If there is evidence of inflammation, a drug with analgesic and anti-inflammatory properties will be used. If the pain has some characteristics of neuropathic pain, one of the drugs commonly used for that type of pain is used. If pain seems to occur when the child is sitting for a lengthy period of time, alterations may be made to the number of hours he is sitting. Both the intended actions and the side effects of each intervention (drug or a behavioural intervention) has to be considered.

Optimally, one treatment is given at a time and the effect of the treatment observed over a few days. If the pain is alleviated, the treatment may be withdrawn for a short period of time to see if the pain begins to reoccur. This simple reversal design can be a clear indication that the treatment is what is helping the pain.

Research on all types of pain management in children with cognitive impairments is seriously lacking. Only scattered studies are available. Without any specific research, the pain management approaches shown effective with other children should be tried. However, it is likely that outcomes could be significantly improved if research was able to detail specific strategies that were optimum for children with cognitive impairment.

SUMMARY

As our knowledge grows in pain assessment and management for children with cognitive impairments, it is vital that we use the information we currently have at hand in caring for children now. There is a history of under-treatment of pain in vulnerable groups, with children with cognitive impairments representing only one of these. It can be easy for us to assume differences in pain sensation when no evidence exists, because of our pre-existing beliefs about

pain, about the relation between pain and intellectual functioning, and about the pain tolerance of those who may express pain differently from us.

Undoubtedly, pain is a subjective experience. It may vary due to many factors, such as age, gender and culture, and sensitivity and tolerance can be subject to psycho-emotional context. The suffering associated with pain is also subjective. It is the very subjectivity of pain and suffering that raises obstacles for us. Empathy is fundamental to evaluating the pain of another, and this requires identification with the person suffering pain when we have no objective source of information. However it is difficult to identify with those who we see as “different”, leaving us feeling helpless and inadequate to the task. Thus, we may develop mechanisms to deal with our frustrations when working with those who we view as “impaired”. This can include minimizing their experience, or its impact on their lives. These attitudes are as likely to influence pain management as they are other aspects of care with these groups.

A child with cognitive impairments can elicit our pity and charity. On the other hand, humans still display a general tendency to “blame the victim” in our efforts to make sense of suffering. These two feelings can lead to an ambivalence that can also interfere with good pain management, and may be one reason that recognition of pain may be delayed for this group. This highlights the fact that any tool is dependent upon the hand which holds it.

In all, there is little doubt that objective assessment is the best guarantee that treatment will be administered and will be effective. Objective tools can help to minimize our biases, to detect patterns in pain and to adapt treatment on an ongoing basis. It also makes it possible to communicate reliable information with all involved in the care of a child and helps professionals to become proficient in pain assessment and management strategies. Objective assessment also allows us to collect information across children to improve our understanding of specific pain problems.

The insistence that children with cognitive impairments are idiosyncratic underlies not only resistance to use of structured pain assessment, but also arguments that pain treatments will not be effective for them. When benefits outweigh potential harm, we have little evidence that treatments that are effective for specific pain problems in typical children will not be so in those with cognitive impairments.

Our challenge at this juncture is to achieve balance. To accept and be sensitive to the uniqueness of each child and their personal experience of pain, while pressing on in our efforts to validate objective pain assessment tools that will aid in consistent high-quality diagnosis, to gather evidence regarding the effectiveness of pain management strategies for these very complex children, and to promote the use of scientifically tested methods in all aspects of pain care.

REFERENCE LIST

- Adhami, E. J. & Cancio-Babu, C. V. (2003). Anaesthesia in a child with Sotos syndrome. *Paediatr Anaesth.*, 13, 835-840.
- Allison, P. J. & Lawrence, H. P. (2004). A paired comparison of dental care in Canadians with Down syndrome and their siblings without Down syndrome. *Community Dentistry and Oral Epidemiology*, 32, 99-106.
- Arvio, M. & Sillanpaa, M. (2003). Prevalence, aetiology and comorbidity of severe and profound intellectual disability in Finland. *Journal of Intellectual Disability Research*, 47, 108-112.
- Benedetti, F., Arduino, C., Vighetti, S., Asteggiano, G., Tarenzi, L., & Rainero, I. (2004). Pain reactivity in Alzheimer patients with different degrees of cognitive impairment and brain electrical activity deterioration. *Pain*, 111, 22-29.
- Benini, F., Trapanotto, M., Gobber, D., Agosto, C., Carli, G., Drigo, P. et al. (2004). Evaluating pain induced by venipuncture in pediatric patients with developmental delay. *Clin.J Pain*, 20, 156-163.
- Biersdorff, K. K. (1991). Pain insensitivity and indifference: alternative explanations for some medical catastrophes. *Mental Retardation*, 29, 359-362.

- Biersdorff, K. K. (1994). Incidence of significantly altered pain experience among individuals with developmental disabilities. *American Journal on Mental Retardation*, 98, 619-631.
- Birmingham, P. K., Wheeler, M., Suresh, S., Dsida, R. M., Rae, B. R., Obrecht, J. et al. (2003). Patient-controlled epidural analgesia in children: can they do it? *Anesthesia and Analgesia*, 96, 686-91, table.
- Bosch, J. J. (2002). Use of directed history and behavioral indicators in the assessment of the child with a developmental disability. *J.Pediatr.Health Care*, 16, 170-179.
- Braden, K., Swanson, S., & Di Scala, C. (2003). Injuries to children who had preinjury cognitive impairment: a 10-year retrospective review. *Arch.Pediatr.Adolesc Med*, 157, 336-340.
- Breau, L. M., Camfield, C., Symon, D. N., Bodfish, J. W., McKay, A., Finley, G. A. et al. (2003a). Pain and self-injurious behaviour in neurologically impaired children. *Journal of Pediatrics*, 142, 498-503.
- Breau, L. M., Camfield, C. S., McGrath, P. J., & Finley, G. A. (2003b). The incidence of pain in children with severe cognitive impairments. *Archives of Pediatrics & Adolescent Medicine*, 157, 1226.
- Breau, L. M., Camfield, C. S., McGrath, P. J., & Finley, G. A. (2004a). Risk factors for pain in children with severe cognitive impairments. *Developmental Medicine and Child Neurology*, 46, 364-371.
- Breau, L. M., Camfield, C. S., McGrath, P. J., Rosmus, C., & Finley, G. A. (2001). Measuring Pain Accurately in Children with Cognitive Impairments: Refinement of a Caregiver Scale. *Journal of Pediatrics*, 138, 721-727.
- Breau, L. M., Finley, G. A., McGrath, P. J., & Camfield, C. S. (2002). Validation of the Non-Communicating Children's Pain Checklist - Postoperative Version. *Anesthesiology*, 96, 528-535.
- Breau, L. M., MacLaren, J., McGrath, P. J., Camfield, C. S., & Finley, G. A. (2003). Caregivers' beliefs regarding pain in children with cognitive impairment: relation between pain sensation and reaction increases with severity of impairment. *Clinical Journal of Pain*, 19, 335-344.
- Breau, L. M., McGrath, P. J., Camfield, C., & Finley, G. A. (2002). Psychometric Properties of the Non-communicating Children's Pain Checklist-Revised. *Pain*, 99, 349-357.
- Breau, L. M., McGrath, P. J., Camfield, C., Rosmus, C., & Finley, G. A. (2000). Preliminary validation of an observational pain checklist for persons with cognitive impairments and inability to communicate verbally. *Developmental Medicine and Child Neurology*, 42, 609-616.
- Breau, L. M., McGrath, P. J., Craig, K. D., Santor, D., Cassidy, K. L., & Reid, G. J. (2001). Facial expression of children receiving immunizations: A principal components analysis of the Child Facial Coding System. *Clinical Journal of Pain*, 17, 178-186.
- Breau, L. M., McGrath, P. J., Stevens, B., Beyene, J., Camfield, C., Finley, G. A. et al. Healthcare Professionals' Beliefs Regarding the Pain of Infants at Risk for Neurological Impairment: A Survey. *Clinical Journal of Pain*, (in press).
- Breau, L. M., Stevens, B., McGrath, P. J., Beyene, J., Camfield, C. S., Finley, G. A. et al. (2004b). Healthcare professionals' perception of pain experienced by infants at risk for neurological impairment. *BMC Pediatrics*, 4.
- Carter, B., McArthur, E., & Cunliffe, M. (2002). Dealing with uncertainty: parental assessment of pain in their children with profound special needs. *J Adv Nurs*, 38, 449-457.
- Chambers, C. T., Cassidy, K. L., McGrath, P. J., Gilbert, C. A., & Craig, K. D. (1996). *Child Facial Coding System: A Manual*. Dalhousie University and University of British Columbia.
- Choo, S. A., McGrath, P. J., Finley, G. A., Camfield, C., & Breau, L. M. (2001). Causes of Pain in Non-Verbal Cognitively Impaired Children - A Delphi Study.
- Ref Type: Unpublished Work
- Cohen, L. L., Blount, R. L., & Panopoulos, G. (1997). Nurse coaching and cartoon distraction: an effective and practical intervention to reduce child, parent, and nurse distress during immunizations. *Journal of Pediatric Psychology*, 22, 355-370.
- Collignon, P. & Giusiano, B. (2001). Validation of a pain evaluation scale for patients with severe cerebral palsy. *European Journal of Pain*, 5, 433-442.
- Collignon, P., Giusiano, B., Porsmoguer, E., Jimeno, M. E., & Combe, J. C. (1995). Difficultés du diagnostic de la douleur chez l'enfant polyhandicapé. *Annals of Pediatrics*, 42, 123-126.
- Collignon, P., Porsmoguer, E., Behar, M., Combe, J. C., & Perrin, C. (1992). L'automutilation: expression de la douleur chez le sujet déficient mental profond. In *La Douleur de l'Enfant Quelles Responses?* (pp. 15-21). Paris: UNESCO.

- Courreges, P., Nieuviarts, R., & Lecoutre, D. (2003). Anaesthetic management for Edward's syndrome. *Paediatr Anaesth.*, *13*, 267-269.
- Critchley, L. A., Gin, T., & Stuart, J. C. (1995). Anaesthesia in an infant with Rubinstein-Taybi syndrome. *Anaesthesia*, *50*, 37-38.
- Defrin, R., Pick, C. G., Peretz, C., & Carmeli, E. (2004). A quantitative somatosensory testing of pain threshold in individuals with mental retardation. *Pain*, *108*, 58-66.
- Ehde, D. M., Jensen, M. P., Engel, J. M., Turner, J. A., Hoffman, A. J., & Cardenas, D. D. (2003). Chronic pain secondary to disability: a review. *Clinical Journal of Pain*, *19*, 3-17.
- Eland, J. M. (1985). The child who is hurting. *Semin Oncol Nurs*, *1*, 116-122.
- Fanurik, D., Koh, J. L., Harrison, D., & Conrad, T. M. (1996). Children with cognitive impairment: parent report of pain and coping. Proceedings of the 8th World Congress on Pain, Progress in Pain Research and Management , 299. Ref Type: Abstract
- Fanurik, D., Koh, J. L., Harrison, R. D., Conrad, T. M., & Tomerlin, C. (1998). Pain assessment in children with cognitive impairment: An exploration of self-report skills. *Clinical Nursing Research*, *7*, 103-124.
- Fanurik, D., Koh, J. L., Schmitz, M. L., Harrison, R. D., & Conrad, T. M. (1999a). Children with cognitive impairment: Parent report of pain and coping. *Journal of Developmental and Behavioral Pediatrics*, *20*, 228-234.
- Fanurik, D., Koh, J. L., Schmitz, M. L., Harrison, R. D., Roberson, P. K., & Killebrew, P. (1999b). Pain assessment and treatment in children with cognitive impairment: A survey of nurses' and physicians' beliefs. *Clinical Journal of Pain*, *15*, 304-312.
- Fernhall, B. & Otterstetter, M. (2003). Attenuated responses to sympathoexcitation in individuals with Down syndrome. *J Appl Physiol*, *94*, 2158-2165.
- Finley, G. A., McGrath, P. J., Forward, S. P., McNeill, G., & Fitzgerald, P. (1996). Parents' management of children's pain following 'minor' surgery. *Pain*, *64*, 83-87.
- Gillberg, C., Terenius, L., & Lonnerholm, G. (1985). Endorphine activity in childhood psychosis. Spinal fluid levels in 24 cases. *Archives of General Psychiatry*, *42*, 780-783.
- Giusiano, B., Jimeno, M. T., Collignon, P., & Chau, Y. (1995). Utilization of a neural network in the elaboration of an evaluation scale for pain in cerebral palsy. *Methods of Information in Medicine*, *34*, 498-502.
- Hadden, K. L. & von Baeyer, C. L. (2001). Measuring Pain In Children with Cerebral Palsy During Home Stretching Exercises: Facial Actions and Global Ratings. Canadian Pain Society, Montreal, May. *Pain Research and Management*, *6*(Supp A), 39A-40A (abstract).
- Hadden, K. L. & von Baeyer, C. L. (2002). Pain in children with cerebral palsy: common triggers and expressive behaviors. *Pain*, *99*, 281-288.
- Halperin, S. A., McGrath, P., Smith, B., & Houston, T. (2000). Lidocaine-prilocaine patch decreases the pain associated with the subcutaneous administration of measles-mumps-rubella vaccine but does not adversely affect the antibody response. *J.Pediatr.*, *136*, 789-794.
- Hennequin, M., Faulks, D., & Roux, D. (2000). Accuracy of estimation of dental treatment need in special care patients. *J Dent.*, *28*, 131-136.
- Hennequin, M., Morin, C., & Feine, J. S. (2000). Pain expression and stimulus localisation in individuals with Down's syndrome. *Lancet*, *356*, 1882-1887.
- Hicks, C. L., von Baeyer, C. L., Spafford, P. A., van, K., I, & Goodenough, B. (2001). The Faces Pain Scale - Revised: toward a common metric in pediatric pain measurement. *Pain*, *93*, 173-183.
- Hunt, A. & Burne, R. (1995). Medical and nursing problems of children with neurodegenerative disease. *Palliative Medicine*, *9*, 19-26.
- Hunt, A., Goldman, A., Seers, K., Crichton, N., Mastroyannopoulou, K., Moffat, V. et al. (2004). Clinical validation of the paediatric pain profile. *Developmental Medicine and Child Neurology*, *46*, 9-18.
- Hunt, A., Mastroyannopoulou, K., Goldman, A., & Seers, K. (2003). Not knowing--the problem of pain in children with severe neurological impairment. *Int.J.Nurs.Stud.*, *40*, 171-183.
- Iacobucci, T., Galeone, M., & De Francisci, G. (2003). Anaesthetic management of a child with Pallister-Killian syndrome. *Paediatr Anaesth.*, *13*, 457-459.
- Jancar, J. & Speller, C. J. (1994). Fatal intestinal obstruction in the mentally handicapped. *Journal of Intellectual Disability Research*, *38*, 413-422.
- Jay, S. M., Ozolins, M., Elliott, C. H., & Caldwell, S. (1983). Assessment of children's distress during painful medical procedures. *Health Psychology*, *2*, 133-147.

- Konstantareas, M. M. & Homatidis, S. (1987). Ear infections in autistic and normal children. *J. Autism Dev Disord.*, 17, 585-594.
- Leland, N. L., Garrard, J., & Smith, D. K. (1994). Comparison of injuries to children with and without disabilities in a day-care center. *Journal of Developmental and Behavioral Pediatrics*, 15, 402-408.
- Levy, S. E., Souders, M. C., Wray, J., Jawad, A. F., Gallagher, P. R., Coplan, J. et al. (2003). Children with autistic spectrum disorders. I: comparison of placebo and single dose of human synthetic secretin. *Arch. Dis Child*, 88, 731-736.
- Lorenz, J. M., Wooliever, D. E., Jetton, J. R., & Paneth, N. (1998). A quantitative review of mortality and developmental disability in extremely premature newborns. *Archives of Pediatrics & Adolescent Medicine*, 152, 425-435.
- Lu, D. P. (1981). Clinical investigation of relative indifference to pain among adolescent mental retardates. *ASDC Journal of Dentistry for Children*, 48, 285-288.
- Malviya, S., Voepel-Lewis, T., Tait, A. R., Merkel, S., Lauer, A., Munro, H. et al. (2001). Pain management in children with and without cognitive impairment following spine fusion surgery. *Paediatric Anaesthesia*, 11, 453-458.
- McDonald, A. J. & Cooper, M. G. (2001). Patient-controlled analgesia: an appropriate method of pain control in children. *Paediatr Drugs*, 3, 273-284.
- McGrath, P. J., Rosmus, C., Camfield, C., Campbell, M. A., & Hennigar, A. W. (1998). Behaviours caregivers use to determine pain in non-verbal, cognitively impaired individuals. *Developmental Medicine and Child Neurology*, 40, 340-343.
- Mercer, K. & Glenn, S. (2004). The expression of pain in infants with developmental delays. *Child Care Health Dev.*, 30, 353-360.
- Mette, F. & Abittan, J. (1988). Essais d'évaluation de la douleur chez le polyhandicapé. *Annales Kinésithérapie*, 15, 101-104.
- Nader, R., Oberlander, T. F., Chambers, C. T., & Craig, K. D. (2004). Expression of pain in children with autism. *Clin. J Pain*, 20, 88-97.
- Nordin, V. & Gillberg, C. (1996). Autism spectrum disorders in children with physical or mental disability or both. I: Clinical and epidemiological aspects. *Dev Med Child Neurol*, 38, 297-313.
- Oberlander, T. F., Gilbert, C. A., Chambers, C. T., O'Donnell, M. E., & Craig, K. D. (1999). Biobehavioral responses to acute pain in adolescents with a significant neurologic impairment. *Clinical Journal of Pain*, 15, 201-209.
- Porter, F. L., Malhotra, K. M., Wolf, C. M., Morris, J. C., Miller, J. P., & Smith, M. C. (1996). Dementia and response to pain in the elderly. *Pain*, 68, 413-421.
- Rainero, I., Vighetti, S., Bergamasco, B., Pinessi, L., & Benedetti, F. (2000). Autonomic responses and pain perception in Alzheimer's disease. *European Journal of Pain*, 4, 267-274.
- Reynell, J. K. (1965). Post-operative disturbances observed in children with cerebral palsy. *Developmental Medicine and Child Neurology*, 7, 360-376.
- Sandman, C. A., Barron, J., Chicz-DeMet, A., & DeMet, E. M. (1990). Plasma B-Endorphin levels in patients with self-injurious behavior and stereotypy. *American Journal on Mental Retardation*, 95, 84-92.
- Shenkman, Z., Krichevski, I., Elpeleg, O. N., Joseph, A., & Kadari, A. (1997). Anaesthetic management of a patient with Leigh's syndrome. *Can. J Anaesth.*, 44, 1091-1095.
- Soetenga, D., Pellino, T. A., & Frank, J. (1999). Assessment of the validity and reliability of the University of Wisconsin Children's Hospital pain scale for preverbal and nonverbal children. *Pediatric Nursing*, 25, 670-676.
- Solodiuk, J. & Curley, M. A. (2003). Pain assessment in nonverbal children with severe cognitive impairments: the Individualized Numeric Rating Scale (INRS). *Journal of Pediatric Nursing*, 18, 295-299.
- Souders, M. C., Freeman, K. G., DePaul, D., & Levy, S. E. (2002). Caring for children and adolescents with autism who require challenging procedures. *Pediatric Nursing*, 28, 555-562.
- Stallard, P., Williams, L., Lenton, S., & Velleman, R. (2001). Pain in cognitively impaired, non-communicating children. *Archives of Disease in Childhood*, 85, 460-462.
- Stallard, P., Williams, L., Velleman, R., Lenton, S., McGrath, P. J., & Taylor, G. (2002). The development and evaluation of the pain indicator for communicatively impaired children (PICIC). *Pain*, 1-2, 149.

- Stephenson, J. R. & Dowrick, M. (2000). Parent priorities in communication intervention for young students with severe disabilities. *Education and Training in Mental Retardation and Developmental Disabilities, 35*, 25-35.
- Stevens, B., McGrath, P. J., Gibbins, S., Beyenne, J., Breau, L. M., Camfield, C. et al. (2003). Procedural pain in neonates at risk for neurological impairment. *Pain, 105*, 27-35.
- Terstegen, C., Koot, H. M., de Boer, J. B., & Tibboel, D. (2003). Measuring pain in children with cognitive impairment: pain response to surgical procedures. *Pain, 103*, 187-198.
- Tracy, J. M. & Wallace, R. (2001). Presentations of physical illness in people with developmental disability: the example of gastro-oesophageal reflux. *Med.J Aust., 175*, 109-111.
- Voepel-Lewis, T., Merkel, S., Tait, A. R., Trzcinka, A., & Malviya, S. (2002). The reliability and validity of the Face, Legs, Activity, CRY, Consolability observational tool as a measure of pain in children with cognitive impairment. *Anesthesia and Analgesia, 95*, 1224-1229.
- Wong, D. L. & Baker, C. M. (1988). Pain in children: comparison of assessment scales. *Pediatric Nursing, 14*, 9-17.
- Zabalía, M., Jacquet D. & Breau L.M. (2005) rôle du niveau verbal sur l'expression et l'évaluation de la douleur chez des sujets déficients intellectuels, *Doul. et Analg. 2*, 65-70.

Table 1: Items of three observational pain tools that provide clinical cut-off scores for pain

NCCPC-R	PPP	DESS
Moaning , whining, whimpering (Fairly soft)	Cries / moans / groans / screams or whimpers	Moaning or inaudible cries (cries with manipulation or spontaneously, in an irregular or continuous way)
Crying (Moderately loud)		Crying (cries with or without tears)
Screaming/yelling (Very loud)		
A specific sound or word for pain (for example: a word, cry, or type of laugh)		
Eating less, not interested in food	Is reluctant to eat / difficult to feed	
Increase in sleep		
Decrease in sleep	Has disturbed sleep	
Not co-operating, cranky, irritable, unhappy	1)Is cheerful (reverse scored) 2) Is hard to console or comfort	Ability to communicate with the nurse (by searching, expressions or babbles, spontaneously or when being solicited)
Less interaction with others, withdrawn	1) Is sociable or responsive (reverse scored) 2) Appears withdrawn or	Spontaneous interest for the surroundings (negatively rated)

NCCPC-R	PPP	DESS
	depressed	
Seeking comfort or physical closeness		
Being difficult to distract, not able to satisfy or pacify		
A furrowed brow	Frowns / has furrowed brow / looks worried	
A change in eyes, including: squinching of eyes, eyes opened wide; eyes frowning	1) Grimaces / screws up face / screws up eyes 2) Looks frightened (with eyes wide open)	Painful expression (the face shows pain; a paradoxical laugh can correspond to a painful rictus)
Turning down of mouth, not smiling		
Lips puckering up, tight, pouting or quivering		
Clenching or grinding teeth, chewing or thrusting tongue out	Grinds teeth or makes mouthing movements	
Not moving, less active, quiet		
Jumping around, agitated, fidgety	1) Is restless / agitated or distressed 2) Has involuntary or stereotypical movements / is jumpy / startles or	Increase in spontaneous movement (voluntary motricity or not, coordinate or not, choreoathetotic)

NCCPC-R	PPP	DESS
	has seizures	movements of limbs or head)
Floppy		
Stiff, spastic, tense, rigid	Tenses / stiffens or spasms	Aggravation of tonic troubles (increase in stiffness, tremulations, hypertonic spasms)
Gesturing to or touching part of the body that hurt	Tends to touch or rub particular areas	
Protecting, favouring or guarding part of the body that hurts	Resists being moved	Protection of painful areas (protects the area supposed painful with his/her hand in order to avoid contact)
Flinching or moving the body part away, being sensitive to touch	Pulls away or flinches when touched	Coordinated defensive reaction or equivalent on examination of an area supposed painful (grazing, touching or mobilization induces a coordinated bodily reaction or equivalent that we can interpret as a defensive reaction)
Moving the body in specific way to show pain (e.g. head back, arms down, curls up etc.)	<ol style="list-style-type: none"> 1) Flexes inward or draws legs up towards chest 2) Twists and turns / tosses 	Spontaneous antalgic position (search for an unusual position that calms) or placed in antalgic

NCCPC-R	PPP	DESS
	head / writhes or arches back	position by nurse)
Shivering		
Change in colour, pallor		
Sweating, perspiring		
Tears		
Sharp intake of breath, gasping		
Breath holding		
	Self-harms, e.g. biting self or banging head	
