THE INITIAL VALIDATION OF A NON-VOCAL, MULTIDIMENSIONAL PAIN ASSESSMENT INSTRUMENT FOR INDIVIDUALS WITH CHARGE SYNDROME

Kasee K. Stratton

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Dissertation Committee:

Timothy Hartshorne, Ph.D.	Committee Chair
Sharon Bradley-Johnson, Ed.D.	Faculty Member
Kyunghee Han, Ph. D.	Faculty Member
Jude Nicholas, Psy.D.	Faculty Member
November 22, 2011	Date of Defense
Roger Coles, Ed.D.	Dean College of Graduate Studies
April 3, 2012	Approved by the College of Graduate Studies

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ABSTRACT

THE INITIAL VALIDATION OF A NON-VOCAL, MULTIDIMENSIONAL PAIN ASSESSMENT INSTRUMENT FOR INDIVIDUALS WITH CHARGE SYNDROME

by Kasee K. Stratton

The investigation into the identification of pain among individuals with developmental disabilities is a recent area of research, particularly among individuals who are non-vocal. Individuals with CHARGE syndrome, a genetic condition involving a number of physical anomalies present at birth, often have communication impairments. Additionally, these individuals experience considerable pain from multiple medical procedures and other causes. Due to their sometimes limited communication skills and with self-report being the "gold standard" for treatment, individuals with CHARGE are often unable to report their pain using typical behaviors.

Further, an investigation by Stratton and Hartshorne (2010) reported that individuals with CHARGE experience acute and chronic pain; however, current non-vocal pain instruments have been found to present with some items that do not discriminate pain for individuals with CHARGE and have missed opportunities to improve reliability and validity for individuals with CHARGE. The purpose of the present project was to determine the validation and psychometric properties (reliability and validity) of a non-vocal pain assessment instrument, designed for individuals with CHARGE. The CHARGE Non-Vocal Pain Assessment (CNVPA) was constructed based on pain behaviors described by parents and from items discriminating pain on two established non-vocal pain assessments in Stratton and Hartshorne's (2010) investigation.

Results found that the CHARGE Non-Vocal Pain Assessment (CNVPA) discriminated between when children were in pain from no-pain episodes. The CNVPA is a reliable and valid measure for individuals with CHARGE when conducted in naturalistic settings (e.g. home) and completed by parents and familiar others. This assessment was also found to be a useful and socially valid instrument for individuals with CHARGE. Some variations in pain behaviors were noted based on the age of the individuals rated with the CNVPA; however, further investigation is needed.

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CHAPTER I

INTRODUCTION

Pain and Disability

Pain serves as an essential protective function that prompts individuals to seek treatment (Turk & Melzack, 2001). To seek treatment, one must first identify pain. Medical professionals and the like have considered the "gold standard" of pain identification and treatment to be patient self-report (Bottos & Chambers, 2006). When a patient self-reports, they are able to vocally verbalize their pain. In addition, individuals can also provide an explanation of where the pain occurs, the duration of time they have been experiencing pain, and the intensity of pain/discomfort. Individuals with communication impairments or limitations and those with developmental disabilities may not be able to vocally self-report their pain experiences, leaving them vulnerable to no treatment or inconsistent pain management. As a result, pain in children has become a recent subject of research, particularly for children with developmental or acquired disabilities.

In addition to having received little research attention, individuals with developmental disabilities and limited communication have also been portrayed as having a higher threshold for pain than the general population or some have suggested that these individuals did not experience pain at all (Sobsey, 2006). Little research supported such notions, however, as past pain research investigations excluded individuals with developmental disabilities for a variety of reasons: measurement only by self-report, the possibility that pain experiences differed from the typically-developing population, or that their advanced medical conditions lead to complicated pain assessments (Breau,

McGrath, & Zabalia, 2006). More recently researchers found that the pain experience of those with developmental disabilities is likely very similar to that of the general population; however, these individuals may communicate pain in a different way (Oberlander & Symons, 2006). No evidence exists to suggest that individuals with developmental or acquired disabilities are not insensitive or indifferent to pain.

Pain is significant in the lives of children and adults with developmental disabilities. First, individuals with developmental disabilities are at an increased risk for experiencing more frequent severe pain due to physical impairments and managing their conditions with medical procedures (Breau, Camfield, McGrath, & Finley, 2003; Stallard, Williams, Lenton, & Velleman, 2001). Second, individuals are likely to express their pain experiences differently than typically-developing peers, e.g., through their behavior rather than vocally, (Gilbert-MacLeod, Craig, Rocha, & Mathias, 2000) and this may impact the care received and pain management. Additionally, chronic pain and illness can produce traumatic stress which may produce stress disorders, e.g., Posttraumatic Stress Disorder (PTSD) (Kassam-Adams, 2006). Further, untreated pain and decreased pain management can significantly impact behavior, relationships and emotional attachment, adaptive functioning, educational experiences, and can produce anxiety and depression (Breau, Camfield, McGrath, & Finley, 2007; Kennedy & O'Reilly, 2006; Palermo, 2000; Porter, Davis, & Keefe, 2007; Power, Heathfield, McGoey & Blum, 1999).

Individuals with genetic anomalies are also at increased risk for higher pain and limited self-report. Additionally, these individuals can be significantly impacted by the negative consequences of pain on behavior.

CHARGE syndrome is a genetic condition involving a large number of physical anomalies present at birth, including heart defects and deaf-blindness (Pagon, Graham, Zonana, & Yong, 1981). Additional anomalies, including major and minor diagnostic characteristics, are presented in Table 1 and 2 (Hartshorne, Hefner, Davenport, & Thelin, 2010).

Table 1. *Major Diagnostic Characteristics of CHARGE Syndrome*

Characteristic	**Manifestations**	**Frequency**
Coloboma of the eye	Coloboma of the iris, retina, choroid, disc; microphthalmos	80 – 90%
Choanal atresia or stenosis [1, 2]	Unilateral/bilateral: bony/membranous, atresia/stenosis	50 – 60%
Cranial nerve dysfunction or anomaly	I: Hyposmia or anosmia/arhinencephaly	>90%
	VII: Facial palsy (unilateral or bilateral)	40%
	VIII: Hypoplasia of auditory nerve	40%
	IX/X: Swallowing problems with aspiration	70 - 90%
Characteristic CHARGE outer ear	Short, wide ear with little or no lobe. A "snipped off" helix, prominent antihelix which is often discontinuous with tragus, triangular concha, decreased cartilage, often protrude, usually asymmetric	>50%
Characteristic CHARGE middle or inner ear	Ossicular malformations	>80%
	Mondini defect of the cochlea	>80%
	Absent or hypoplastic semicircular canals	>90%

1. Choanae are passages in the back of the nose which are blocked (atretic) or narrowed (stenotic)
2. Cleft palate may substitute for this characteristic in some cases

From *CHARGE Syndrome* by Hartshorne, T.S., Hefner, M., Davenport, S., & Thelin, J.W. (eds.).

Table 2. *Minor Diagnostic Characteristics of CHARGE Syndrome*

Characteristic	Manifestations	Frequency
Genital hypoplasia	*Males*: Micropenis, cryptorchidism	50%
	Females: Hypoplastic labia	50%
	Both: Delayed puberty	>50%, 90% in males
Cardiovascular malformation	Especially conotruncal defects (e.g., tetralogy of Fallot), aortic arch anomalies	75-85%
Growth deficiency	Short stature	70%
	Growth hormone deficiency	15%
Orofacial cleft	Cleft lip and/or palate	15-20%
Tracheoesophageal (T-E) fistula	T-E defects of all types	15-20%
Renal anomalies	Ectopic or solitary or duplex kidney, UPJ obstruction, reflux, hydronephrosis	30-40%
Distinctive facial features	Square face with broad prominent forehead, prominent nasal bridge and columella, flat midface, small chin which gets larger with age	70-80%
Palmar crease	Hockey-stick palmar crease	50%
CHARGE behavioral profile	OCD or other perseverative behavior	>50%

Individuals with CHARGE syndrome often endure multiple intensive medical procedures resulting in long-term hospital stays and recovery periods (Hartshorne & Cypher, 2004). Many children with CHARGE develop extremely challenging behavior including inattention, obsessive-compulsive behaviors, tantrum outbursts, self-

injury/abuse, aggression toward others, and disruptive vocal and motor responses, i.e., screaming and stereotypy (Hartshorne, Hefner, & Davenport, 2005; Lauger, Cornelius, & Keedy, 2005; van Dijk & de Kort, 2005). Parents and professionals working with children with CHARGE have long suspected that these children experience considerable pain related to some of their behavioral difficulties. Research has shown that challenging behaviors, including aggression and self-injurious behaviors, may be related to an individual's pain experience (Cook, Niven, & Downs, 1999; Symons & Danov, 2005).

Although it is reasonable to assume that children with CHARGE experience pain, and while there is evidence that pain can lead to challenging behaviors, this is difficult to demonstrate with CHARGE because so many children are unable to communicate when they are in pain, and others who can communicate may express pain differently. It is important, therefore, to identify a way to measure the experience of pain for these children.

Stratton and Hartshorne (2010) conducted an investigation to identify pain among children with CHARGE syndrome 18 years of age and under. Results indicated children with CHARGE do experience considerable amounts of pain, although only 75% of parents felt comfortable identifying their child's non-vocal pain behaviors. Two non-vocal pain behavior assessments, the Non-Communicating Children's Pain Checklist-Revised (Breau, McGrath, Finley, & Camfield, 2004) and the Paediatric Pain Profile (Hunt, 2003) (can be found in Appendices A and B, respectively), were used by parents/caregivers (n=53) to identify pain among children with CHARGE, 18 years of age and under. The NCCPC-R and PPP were shown to be useful non-vocal pain

assessments for this population; however, several items did not appear to demonstrate a change in presentation across pain and no-pain measurements and may not be valid for this population. Additionally, parents presented other non-vocal behaviors than those provided on the NCCPC-R and PPP that children with CHARGE display when in pain, e.g., increase in self-injurious and aggressive behaviors. Consequently, these non-vocal pain assessments may not be the most valid or reliable measures of pain for this population.

The present study assessed pain non-vocally among individuals with CHARGE syndrome to examine the validity of an observational, multidimensional, non-vocal pain measure designed specifically for individuals with CHARGE syndrome. This instrument was developed based on the results of Stratton and Hartshorne's (2010) investigation of pain. The purpose of this study was to determine the psychometric properties of this instrument and provide an objective and valid measurement of pain expression in individuals with CHARGE who have limited communication strategies.

CHAPTER II

LITERATURE REVIEW

Pain and Developmental Disabilities

Pain is common to human experience. It is subjective, however, and difficult to directly measure. A common practice, considered the "gold standard" by many in the medical profession (Bottos & Chambers, 2006), is to identify and treat pain through patient self-report. Some individuals, particularly those with developmental disabilities and communication impairments, are unable to self-report their pain experiences. Past research studies have excluded individuals with developmental disabilities for many reasons: use of measurement by self-report, the possibility that pain experiences may differ from typically developing children, or because such advanced medical conditions within this population could complicate assessments (Breau et al., 2006).

Current research on pain provides evidence in support of the perspective that pain is not any less frequent in the lives of those with a developmental or acquired disability; rather, they may alter the way they communicate pain. Those with developmental or acquired disabilities also are not insensitive or indifferent to pain. Limitations may confound their presentation of pain; however, the pain experience may be similar to that of the general population (Oberlander & Symons, 2006).

Gilbert-MacLeod et al. (2000) found children with developmental delays, when compared to non-delayed children, displayed a less intense distress response to everyday pain and failed to respond to the painful event, e.g., falling down. Children without a developmental delay engaged in more help-seeking behavior, e.g., crying. Similarly, infants with developmental delays are less likely to show facial expressions in response

to pain than typically developing children (Mercer & Glenn, 2004). Children in the developmental delay group had received diagnoses of general developmental delay, autism spectrum disorders, Down syndrome, Apert's syndrome, cerebal palsy, and spina bifida.

The lack of response by children with developmental delays may be explained by a socio-communicative deficit. A socio-communicative deficit may result from difficulty learning how to respond to painful experiences. Children with developmental delays may not have acquired help-seeking skills because of limited socialization and because of medical issues early in life that required frequent invasive surgical interventions. Their expression of pain would likely be affected by a reduced opportunity for social interaction and observational learning, due to their health issues and sensory impairments (Craig, 2006).

Recchia (1997) observed 14 toddlers with congenital blindness in a quasi-experimental design to focus on alternative modes of social communication and social referencing from the toddlers to their mothers. All observations of the children and their mothers were completed in the child's home over two 2-hour time periods. Four multi-sensory ambiguous stimuli were presented during typical mother-child play time. After the stimulus was presented, the mother was told to pause briefly to see how her child responded and then to follow through as she would normally behave when introducing something to her child. Stimuli consisted of a stranger standing 3 feet away from the child saying "Hello, (child's name), my name is __." The stranger then approached the child and touched the child's arm, and after 3 seconds, the stranger spoke again. The stranger stimulus was presented every 30 seconds in an attempt to evoke a positive

response. The remaining stimuli were a battery operated toy train with four noise producing buttons, a small accordion, and a manual air pump to blow air near the child's face. The train and accordion were considered to be non-threatening, whereas the stranger and blower were expected to bring about wariness or uncertainly from the child.

The children in this study who used social referencing as an initial strategy were at least 2 years of age; however, these children did not use social referencing consistently. Without vision, the children had inadequate substitutes for accessing information quickly. It was concluded that it takes children with severe visual impairments more time to understand social referencing and to use it effectively, especially if they are less communicative. Should the child also have a hearing impairment, further impacting communication, one can assume it would take much longer to establish social referencing and use it effectively. As a result, this limited ability to interact socially may impact the child's ability to pick up social cues for pain, i.e., facial expressions and interacting with others.

Pain may be significant in children with or without disabilities because of its potential impact on the care received and pain management. Professionals and caretakers who have not developed an accurate procedure to identify pain in individuals with limited communication strategies may miss signs of illness or serious medical complications. Children with intellectual disabilities are at an increased risk for pain going undetected, despite being more likely than typically developing peers to have painful medical conditions or conditions that require painful treatment (Breau et al., 2003). Children with intellectual disabilities more often experience pain that is non-accidental and more chronic and enduring in nature, such as gastrointestinal discomfort, pain from infections,

musculoskeletal pain, and recurrent pain, e.g., ear pain, and pain associated with medical procedures (Breau et al., 2003). In one investigation, caregivers of 94 children (ages 3 to 18 years) with severe cognitive impairments participated in a 4-week study collecting data on the child's frequency, duration, and intensity of pain. Seventy-three children (78%) experienced pain at least once over a 4-week period, and 58 (62%) had non-accidental pain, resulting from more chronic conditions (Breau et al., 2003).

Impact of Pain

Untreated pain and chronic illness can produce chronic stress. Chronic stress is likely to impair daily functioning and can lead to acute stress disorder or Posttraumatic Stress Disorder (PTSD) (Kassam-Adams, 2006). PTSD is a mental condition in which an individual experiences a traumatic event that involves death or threatened death or serious injury. The event must evoke intense fear and helplessness. PTSD symptoms include reliving the event, avoidance/numbing, and feelings of arousal (American Psychiatric Association, 2004).

PTSD is largely associated with war-time traumatic events; however, prolonged and/or unpleasant medical or surgical procedures also can give rise to PTSD. PTSD has been described in studies reviewing cardiac surgery, treatment in intensive care, and human immunodeficiency virus infection (HIV) to name a few (Tedstone & Tarrier, 2003). Patients with cancer also show posttraumatic symptomatology or exhibit PTSD. A review of the literature on cancer patients and PTSD found an average of 5 to 13% of samples reported subclinical PTSD, whereas some reports showed an incidence of PTSD as high as 35% (Kangas, Henry, & Bryant, 2002). Repeated traumatizations from recurring medical or surgical procedures can have pervasive effects on the developing

brain in children and may interfere with neurobiological development. Additionally, this could impact the child's ability to integrate emotional, sensory, and cognitive information, which may impact behavior (van der Kolk, 2005).

When individuals experiencing pain are poorly treated or pain is under-recognized, one's quality of life may be compromised (Oberlander & Symons, 2006). Investigations of chronic pain and its impact on quality of life have largely been conducted among individuals with cerebral palsy. Individuals with cerebral palsy (a life-long neurological disorder that affects body movements and muscle coordination) for example, experience pain from a variety of sources including musculoskeletal deformities, surgical and medical procedures, and rehabilitative interventions. Repeated pain can increase psychological stressors which can negatively impact their quality of life (Engel & Kartin, 2006). Specific quality of life measurements have been developed for this population as a result (Waters, Maher, Salmon, Reddihough, & Boyd, 2005; Waters et al., 2007).

Behavior

Problem behaviors also may increase as a result of pain. Kennedy and O'Reilly (2006) reported approximately 16% of individuals who have a developmental disability engage in some form of problem behavior. Of these problem behaviors, three broad categories exist: aggressive behavior, destructive behavior, and self-injury. Kennedy and O'Reilly theorize that the presence of a health condition may cause discomfort leading to a greater probability of these problem behaviors. Common health conditions such as constipation, gastroesophageal reflux disease, otitis media (middle ear infection), and sleep problems seem to occur more frequently in people with developmental disabilities

(Kennedy & Thompson, 2000). Children with developmental disabilities are more likely than typically developing children to have such painful conditions. If individuals are unable to vocally report the pain or discomfort, problem behaviors could increase as a form of communication.

Symons and Danov (2005) found that self-injury also may increase with pain. They measured ratings of self-injurious behavior and pain using the Self-Injury Trauma Scale (SITS) (Iwata, Pace, Kissel, Nau, & Farber, 1990) and the Non-Communicating Children's Pain Checklist-Revised (NCCPC-R) (Breau et al., 2004) for 2-hour time intervals, three times daily for nine consecutive days. They found time intervals with elevated pain were associated with elevated ratings of self-injurious behaviors.

Symons, Harper, McGrath, Breau, and Bodfish (2009) found additional support for the relationship of chronic self-injurious behaviors and non-vocal pain behaviors. Adult subjects (n=35) with intellectual and developmental disabilities participated in this investigation. Participants were placed into two matched-samples groups. The first group consisted of adults who exhibited significant self-injurious behaviors resulting in a formal treatment plan that occurred on a weekly basis for at least 1-year and produced tissue damage. The control group consisted of participants who were matched for the self-injurious group by level of cognitive impairment (according to the DSM-IV), gender, use of psychotropic medication, and age. Observers completed the Non-Communicating Children's Pain Checklist based on pain-related non-vocal behaviors for one week. Observers were told ratings were used to measure the degree of occurrence of non-verbal behaviors; therefore, they were blind to the purpose of the experiment. Individuals in the self-injurious behavior group had significantly higher total scores than the control group,

indicating that the presence of pain was observed during self-injury. Those individuals with more severe self-injury tended to also exhibit higher overall pain signs. Symons et al. (2009) therefore indicated pain sensation is intact within this population and pain expression non-vocally is amplified for individuals with intellectual disabilities engaging in chronic self-injury.

Pain also might impact attachment and adaptive functioning. The significant associations among various attachment styles and pain need further validation; however, studies have found individuals who report higher levels of pain also score higher on measures of attachment avoidance and anxious attachment (Porter et al., 2007). Parenting an individual with CHARGE is stressful and it has also been shown that parental stress is increased when problem behavior is present. Problem behavior and high parental stress may further impact parent-child attachment for individuals with CHARGE (Wulffaert, Scholte, Dijkxhoorn, Bergman, van Ravenswaaij-Arts, & van Berckelaer-Omnes, 2009).

Additionally, adaptive skills may be impacted by pain. When children are in pain they engage in fewer established adaptive skills (Breau et al., 2007). Breau et al. found that adaptive behaviors impacted by pain included: physical functioning, understanding language (communication), and showing interest in others (socialization). They also found that children with more severe intellectual disability experienced a greater reduction in functioning, between 21 and 29%, when in pain. Furthermore, increased pain intensity resulted in a greater reduction in adaptive skills. Given the fewer adaptive skills practiced, this may also influence long-term functioning.

Assessment

Although research has been limited on pain among individuals with developmental disabilities, in the past decade researchers have begun to explore options for valid assessments to identify pain. One of the first tools designed to measure pain for typically developing children, outside of self-report, was facial reaction to pain. This form of assessment has not been used clinically and needs additional research to determine whether children with impairments show the same facial responses as typically developing children (Breau et al., 2006).

A second type of assessment to identify pain is a multidimensional pain tool. Multidimensional pain tools have been designed for children with developmental or cognitive impairments. Two tools in particular have gained researcher and clinician interest, the Paediatric Pain Profile (PPP) (Hunt, 2003) and the Non-Communicating Children's Pain Checklist (NCCPC) (Breau et al., 2004).

The PPP is a 20-item behavior rating scale designed to assess children without vocal speech who have neurological/cognitive disabilities. Items include Likert scale questions of disturbed sleep, facial expressions, body movements, and self-injurious behavior. Hunt et al. (2004) evaluated the clinical validity of the PPP on 140 children with severe neurological and cognitive impairments who were unable to communicate through speech or augmentative communication. The PPP appears to be a reliable measure with an internal consistency Cronbach's alpha statistic of .83 to .86 for pain measures. Interrater reliability was assessed with 111 children across two different raters and ranged from .74 to .89. To examine the validity of the PPP, 41 children received ratings on the PPP before and four hours following administration of a required analgesic

(pain-reliever). Children had higher PPP mean scores pre-analgesic with falling mean scores during ratings at one, two, and three hours post-analgesic. Further, when children were rated "at their best" and during their "most troublesome pain," PPP scores were significantly higher during pain than the score for "at their best." The psychometric data provide support for the PPP as a reliable and valid measure that can be used in a clinical setting to assess pain in children with severe disabilities. Researchers also state the instrument is sensitive to postoperative pain. PPP assessments were conducted for five days after 30 children had gastrointestinal (n=14) or orthopedic surgery (n=16). Results indicated that average pain scores did not differ across postoperative surgery for all five days; however, the highest pain score was found within the first 24 hours following surgery for 14 children (Hunt et al., 2004).

Hunt et al. (2007) further analyzed the PPP's psychometric properties. Children's behavior in a high-pain group and low-pain group were rated using the PPP within their home or residential setting by parents and researchers during their normal morning routine. Those in the high-pain group were reported by caregivers to have "moderate," "severe," or "very severe" pain that occurred "all the time." Individuals in the low-pain group were found by caregivers to have "no pain" or "mild pain" at "some time each month" or "less than once a month." An independent criterion, saliva cortisol concentration, also was measured to evaluate the PPP. The hypothesis was that saliva cortisol concentrations would positively correlate with PPP scores.

PPP percent scores in the high-pain group were significantly higher than in the low-pain group; further supporting extreme group validity of the PPP, as hypothesized (Hunt et al., 2007). There was a moderate correlation with saliva cortisol levels.

Correlations between PPP percent scores and numerical ratings, made by parents and researchers, (0-10 numerical pain rating scale) ranged from .76 to .90, supporting concurrent validity. Video ratings correlated well with PPP scores and moderately well with numerical ratings by investigators and parents. Intrarater agreement across three raters was found to be .90 for the total PPP score. The authors' attempted to create cut-off scores using video of children from the high pain group that were rated as having moderate or worse pain (an average item score of two or more) from verbal pain ratings made by professionals and parents (video raters) at the time when interrater reliability was assessed. A PPP cut-off score (for each of the moderate to worse pain episodes) of 14 (24.3% or above), resulted in sensitivity at 1.00, indicating how well the PPP identifies those in pain, and specificity at .95, indicating how well the PPP identifies those without pain.

Another extensively researched assessment tool is the Non-Communicating Children's Pain Checklist (NCCPC). McGrath, Rosmus, Camfield, Campbell, and Hennigar (1998) created the initial checklist through parent interviews of children with cerebral palsy and the following seven subscales were then developed: vocal, eating/sleeping, social/personality, facial expression, activity, body and limbs, and physiological. Several subsequent studies provided data in support of the validity of the NCCPC in a home setting to predict behavior during a subsequent painful event, and for use in postoperative pain investigations (Breau et al., 2006). Following these studies the checklist was revised and the current edition is the NCCPC-R, which includes the seven subscales of the original NCCPC. The NCCPC-R has good properties for specificity and sensitivity to pain and scores appear to be consistent over time (Breau et al., 2003). To

examine the NCCPC-R ability to discriminate between pain and no pain over time, subscale scores for two separate pain events were analyzed using a repeated measures ANOVA and the effect of the subscale (seven items showing the highest odds ratio during a pain incident) was significant. The NCCPC-R internal consistency ($\alpha = .79$) approaches acceptable levels for reliability (Breau, McGrath, Camfield, Rosmus, & Finley, 2000). Similar to the PPP, the NCCPC-R has also attempted to create cut-off scores to evaluate sensitivity and specificity. A subtest of seven items was established using the highest percentage of occurrence among items during pain and the highest odds ratios for the NCCPC-R items retrospectively and across time. This subtest was then used for comparison. Results show that 85% of those predicted to have pain did, indicating sensitivity, and 89% of those predicted on the NCCPC-R to not have pain did not have pain, indicating high negative predictive value or specificity (Breau, Camfield, McGrath, Rosmus, & Finley, 2001).

Assessment using the NCCPC-R is based on the child's behavior during a 2-hour time period. Breau, McGrath, Camfield, and Finley (2002) investigated the NCCPC-R's ability to detect pain by comparing scores from pain episode 1 to no-pain episode 1 and those of pain episode 2 to no-pain episode 2. Participants completed 1-week diaries every three months during an 18-month longitudinal study during which pain episodes were recorded. Additionally, during each 1-week diary, caregivers observed their child for 2-hours per day and completed the NCCPC-R. The first pain episode was the first recorded pain incident in the diary, and pain episode 2 was the second pain observation (two days with no pain had to be observed since pain episode 1). Pain episodes included chronic conditions, e.g., gastrointestinal reflux, illness, medical procedures, and injury.

No-pain episodes also were taken from the diaries and the time between no-pain and pain episodes was not found to differ significantly. Between the two episodes all subscales differed significantly, as did the total scores, indicating the ability of the NCCPC-R to discriminate between the presence and absence of pain and further supporting the checklist's discriminant validity. All items also differed significantly between pain and no-pain measures. A Cronbach's alpha of .93 was found for the internal reliability for pain episode 1 and pain episode 2, suggesting the items on the NCCPC-R measure the same construct. Pearson correlations between caregivers' numerical pain ratings and NCCPC-R total score were significantly correlated and several subscales also were significantly correlated. Consistency of behavior also was found across pain episodes and baseline observations. The NCCPC-R serves as a functional measure of pain, because the total scores were significantly correlated with caregivers' numerical ratings of their children's pain intensity. Overall, reliability and validity data appear promising and cutoff scores for pain have been established to make use of the NCCPC-R clinically. A total score of 7 or higher was found to have good sensitivity (84%) and good specificity (65-77%) for detecting pain when pain ratings were compared to no-pain ratings on the NCCPC-R using a ROC curve (Breau et al., 2002).

Defrin, Lotan, and Pick (2006) investigated how acute pain from influenza vaccinations impacted the behavior of individuals with cognitive impairment across levels of impairment (mild, moderate, severe, and profound). Two coding systems, the NCCPC-R and the Facial Action Coding System (FACS) (Ekman & Friesen, 1978), were used to assess behavior across levels of impairment and a control group of individuals with average intelligence. FACS is an instrument used to identify and measure facial

behaviors based on the muscles that produce them. During vaccination good α-values were found for both the FACS and the NCCPC-R (.78 and .84, respectively), supporting internal reliability for these instruments. A main effect of time from baseline to vaccination was found on the NCCPC-R for all groups with a significant increase in scores during the vaccination, indicating the NCCPC-R is sensitive to changes in pain behavior across all groups. Individuals with severe-profound cognitive impairment did not demonstrate an increase in FACS scores during the vaccination; however, they did demonstrate an increase in NCCPC-R ratings, as they exhibited more body movement. Body movements are not measured on the FACS. The NCCPC-R is, therefore, more suitable for individuals with all levels of cognitive impairment. Agreement in NCCPC-R ratings also was found between observers (.80): a caretaker and an independent observer. The main effect of the coder was not significant for the NCCPC-R, nor was the interaction between items and the coder, indicating the scores of the coders were not significantly different with respect to the items and scores chosen. This indicates caregivers can provide reliable scores of behavior during acute pain and presents evidence of inter-scorer reliability. Similar results for inter-scorer reliability have been found in previous studies using the NCCPC-R (Breau et al., 2002).

The NCCPC was designed for use with children. Bodfish, Harper, Deacon, and Symons (2001) developed an adult adaptation of the NCCPC, the Pain and Discomfort Scale (PADS). PADS is designed to assess pain in adults who do not use verbal language or who are cognitively impaired. This 18-item instrument focuses on observable behaviors in several areas as indicators of pain: vocal, mood/interaction, facial expression, body and limbs, and physiological. A four-point Likert scale was added to

this measure to score the severity/intensity of each item. PADS uses a standardized training, examination, and scoring procedure to ensure all body areas are observed for possible pain signs (Bodfish et al., 2001).

Phan, Edwards, and Robinson (2005) evaluated the sensitivity of the PADS in patients with cognitive impairments and communication deficits by detecting pain and discomfort during dental examinations and polishing. All 28 participants had a history of dental examinations. Baseline observations were conducted 30 minutes prior to the appointment in the patient's home and in the waiting room at the dental clinic. After the participant was seated in the dental chair and the dentist/hygienist was present, a third observation occurred. A fourth observation was conducted after the scaling and polishing were complete and lastly within 30 minutes after the participant arrived home. Each observation was conducted over a 2-minute period with an average length of 6 minutes per observation. Observations were also conducted during the dental procedures that were expected to produce acute discomfort or pain. Scores on the PADS during the dental procedure were significantly greater than scores during baseline observations in the home, waiting room, and dental chair prior to the procedure. Scores during the procedure also were significantly greater than scores obtained post-procedure. Phan et al. (2005) concluded the PADS had utility in detecting pain and discomfort during noxious procedures. Although the scale was functionally sensitive to behaviors related to pain, it lacked specificity, because results could not distinguish between moderate discomfort and anxiety. Because of the use of a multiple-baseline design, however, controls were in place to account for the reactions observed. Furthermore, distinctions between anxiety

and pain have been difficult to determine even among patients who are verbal and who possess higher cognitive skills (Sokol, Sokol, & Sokol, 1985).

Lotan, Moe-Nilssen, Ljunggren, and Strand (2010) also extended the NCCPC-R to create an 18-item version titled the Non-Communicating Adult Pain Checklist (NCAPC) for adults with intellectual and developmental disabilities. The NCAPC follows the same Likert-response format as the NCCPC-R for presenting levels of behavior: 0- not at all, 1- just a little, 2-fairly often, and 3-very often. The scale also includes subscales similar to the NCCPC-R: vocal and emotional reactions, facial expression, body language, protective reactions, and physiological reactions. Lotan et al.'s data collection is limited to adults (age range 21-62, *M*= 40.7, *SD*= 14.6) who have an intellectual or developmental disability and who reside in a residential setting in Israel (*n*=59).

Participants were observed during a dental hygiene treatment for two 5-minute periods, while receiving an influenza vaccination (previously video-recorded), and during two non-pain assessments. The first non-pain assessment was conducted while the individual was seated in the residential living room for 2 minutes. The second non-pain assessment occurred in the waiting room just prior to the dental treatment for two 5-minute observation periods. A significant difference was not found between non-pain assessments; however, when comparing pain assessments to non-pain measurements it was found that the mean NCAPC sum scores were significantly higher during pain situations. Participants mean total scores on the NCAPC were also significantly higher during the dental treatment than during the influenza injection, as hypothesized by the authors. A Pearson correlation (r =.88) suggests that during the dental treatment and

influenza injection the participants responded in a similar manner. In addition, results were found to be useful for adults across all levels of intellectual impairment (profound, severe, moderate, and mild) (Lotan et al., 2010).

Certainly, individuals may exhibit several types of non-vocal behaviors to communicate pain. Such pain behaviors may include increased flexion or extension of the limbs, self-harm, hitting self, putting hands in mouth, grunting, hypo-sensitivity to sound or touch due to sensory overload, rocking or head banging, or additional changes from normal routines. Parents may be the best resource for identifying abnormal behavior in their child, particularly when children are experiencing pain (Twycross, Mayfield, & Savory, 1999). Additionally, multidimensional assessment tools like the PPP and NCCPC-R provide promise in aiding parents, physicians, and others in the identification of pain behavior in children. Additionally, these behavior rating scales describe the intensity and frequency of behavior, an advantage over other observational measures (von Baeyer & Spagrud, 2007). Instruments such as the adult version (NCAPC) of the NCCPC-R, show promise for use with adults who have limited communication strategies; however, further studies are needed to understand non-vocal pain behaviors displayed by adults.

CHARGE Syndrome

CHARGE is a complex genetic syndrome based on several common birth defects/features varying from child to child both medically and physically and following an identifiable pattern genetically. Pagon et al. (1981) recommended the acronym CHARGE based on what they considered to be the most common characteristics: C-Coloboma of the eye, H-Heart defects, A-Atresia of the choanae, R-Retardation of

growth and/or development, G-Genitourinary anomalies, E-Ear abnormalities or deafness.

The acronym is no longer used for diagnosis and revised diagnostic criteria require the presence of three or four major features, common in CHARGE, but uncommon to other syndromes for the diagnosis of CHARGE syndrome: choanal atresia, coloboma, cranial nerve involvements, and characteristic ear abnormalities (Blake et al., 1998). Others have suggested the addition of vestibular dysfunction as a major characteristic (Verloes, 2005). Several minor characteristics such as cleft lip, growth delay, characteristic CHARGE face, and congenital heart defects also are common in CHARGE. Existence of two major characteristics and three minor characteristics suggest a diagnosis of the syndrome (Blake et al., 1998). It is important to note that no single characteristic is universally accepted as a feature of CHARGE and each feature will vary from severe to absent among individuals (Hartshorne et al., 2005). The incidence of CHARGE is generally considered to be approximately 1 in 10,000 births.

It was once believed that all children with CHARGE had intellectual disability. More recent research, however, has found that many individuals with CHARGE display normal intellectual abilities, e.g., individuals may live independently and some complete a college education (Hartshorne et al., 2005; Salem-Hartshorne & Jacob, 2004, 2005). CHARGE involves high to low intellectual functioning and many reports are underestimates of the children's abilities due to problems with communication from examiner to student during testing or lack of professional experience with combined hearing and vision deficits. Multisensory impairment in CHARGE, especially deafblindness (both hearing and vision loss), occurs in 80-90% of cases (Davenport,

Hefner, & Mitchell, 1986) and likely obstructs learning. Deafblindness may result in poor communication strategies; therefore, full classroom inclusion is often not considered. The decision to include a child with CHARGE, however, should be carefully considered based on the unique educational and safety needs of each child. Given appropriate programming, individuals with CHARGE can be successful in each environment. Deafblindness and other sensory impairments may limit the child's access to important learning strategies and access to socially appropriate information. This occurs alongside a host of other medical issues detrimental to the child's ability to communicate and to learn (Hartshorne et al., 2005).

Other complicating issues are damaged or missing semi-circular canals in the ears resulting in balance and coordination problems, in addition to hearing loss. Additionally, individuals with CHARGE may be tube-fed due to difficulty swallowing food as a result of cranial nerve malformations. Numerous surgeries and hospital stays are frequent in the early years of life and continue throughout development. Stratton and Hartshorne (2010) report an average of 12 surgeries for individuals between the ages of 1 to 18 years, with a range of 1 to 47 medical procedures. Examples of such surgeries include several placements of pressure equalization tubes to relieve chronic serous otitis media, also known as chronic ear infections. Because of bony or membranous choanal atresia, airway stabilization is often an immediate need at birth. The posterior choanae must be surgically opened and additional placements of stents are needed to keep the nasal passages open as the child grows, requiring repeated surgical procedures. Tracheotomies are considered necessary for approximately 1 in 20 children with CHARGE to relieve chronic gastroesophageal reflux and aspiration (Blake et al., 1998).

Feeding is an additional concern, with up to 90% of children with CHARGE experiencing some form of a feeding difficulty. As a result, tube feeding or gastrostomies are needed to ensure proper nutrition is obtained. The weakness in the muscles used for sucking, chewing, and swallowing likely result from dysfunction of the cranial nerves V, VII, and IX (Doobelsteyn, Peacocke, Blake, Crist, & Rashid, 2008).

Blake, Hartshorne, Lawand, Dailor, and Thelin (2008) conducted an investigation of cranial nerve anomalies present in CHARGE syndrome. Blake et al. report at least one cranial nerve abnormality present in 92% (*n*= 89) of their sample of individuals with CHARGE (*n*=99), and two or more cranial nerve anomalies in 72% (*n*=71) of the sample. Individuals with CHARGE were shown to have cranial nerve anomalies in nerves V (weak chewing and sucking), VII (facial palsy), VIII (balance/vestibular problems and sensorineural deafness), IX/ X (swallowing problems). Cranial nerve V anomalies were found in over half of the children with CHARGE in this investigation (55.6%). This is an important finding, because migraine headaches have been involved with cranial nerve V (Hargreaves, 2007). Blake, Salem-Hartshorne, Daoud, and Gradstein (2005) linked migraine headaches to pain concerns during adolescence. Further, Stratton and Hartshorne (2010) found migraine headaches to be the most intense painful experience for children (18 years and younger) with CHARGE. Additionally, other trigeminal nerve complications could be a cause for pain among individuals with CHARGE as a result of cranial nerve V anomalies. It has been suspected that all 12 cranial nerves may be impacted in CHARGE, the result of each of these cranial nerve anomalies on pain experiences is also unknown (Blake et al., 2008).

CHARGE Syndrome and Pain Assessment

As previously mentioned, pain assessment can be extremely complicated when individuals are not able to self-report their pain experiences. Communication challenges are a particular concern for CHARGE as many individuals have limited communication due to multisensory impairments (Peltokorpi & Huttunen, 2008; Thelin, Hartshorne, & Hartshorne, 1999). Atypical pain expression is also likely not only as a result of limited communication strategies, but also challenges in sensory integration (Brown, 2005), limited social-emotional experiences, and possible accommodation to and tolerance of chronic pain (Nicholas, 2010). Due to limited vocal communication or the ability to self-report pain experiences, methods must be developed to determine the gestural and bodily expressions of pain for this population, particularly as facial expression scales would be inappropriate due to facial palsy.

Stratton and Hartshorne (2010) conducted the first and only investigation of the identification of pain in children with CHARGE. Parents of 53 children ages 1 to 18 years of age (M=8.87, SD=5.00) participated in the study. Parents completed observational ratings of their child's behavior during a "good day" or when the child was not experiencing pain using the NCCPC-R and PPP. Parents then completed the NCCPC-R and PPP following observations of their child's behavior when the child was experiencing pain, e.g., sinus infection, injury, or migraines. Lastly, parents completed a questionnaire, designed specifically for the study, to identify the non-vocal behaviors each parent identifies/observes when their child experiences pain. Additionally, this questionnaire asked parents to indicate the average duration of pain their child experiences for particular CHARGE characteristics (e.g., gastroesophegeal reflux,

migraines, and sinusitis) and the intensity of this pain on a range from 0 to 4 (0-no hurt, 1-hurts little bit, 2-hurts more, 3-hurts whole lot, and 4-hurts worst).

Children with CHARGE experience a considerable amount of pain. Nearly, all parents (94.3%) indicated their child experiences pain of some degree. Parents rated their child's experience of pain on the following scale: no pain, very little pain, little pain, occasional pain, very frequent pain, and always in pain. The two highest ratings were for "very little pain" and "occasional pain." Even when children with CHARGE are having a "good day," parents indicated mild and moderate pain is still present for over half of the children (52.9%). A majority of parents (75%) indicated they were able to identify pain in their child. On the other hand, 25% of parents of children with CHARGE indicated they were not yet able to identify their child's non-vocal pain behaviors. No significant differences were found for the ability to detect pain based on the age of the child. They also found that no child could vocally or clearly express behaviorally chronic pain, and no parent indicated they could determine if their child was experiencing chronic pain. As a result, when the child is experiencing chronic pain, the percentage of parents able to detect their child's pain is likely to be much less than 75%. This further indicates the need for a non-vocal, multidimensional pain assessment tool.

Behaviors that parents indicated were cues to their child's acute pain fell into similar categories as presented on the NCCPC-R: vocal, social, facial, activity, body and limbs, physiological, and eating/sleeping. However, parents indicated other behaviors that they observed which fall into an additional category not included on the NCCPC-R subscales. This additional category included behavioral challenges, self-injurious behaviors, and dangerous behaviors, as can be seen in Table 3. Children with CHARGE,

according to their parents, appear to display an increase in challenging behaviors when experiencing pain. Because such items are not a part of the PPP or the NCCPC-R, but they are used by parents of children with CHARGE to indicate pain, this suggests that future non-vocal pain assessments should include these items when measuring pain for individuals with CHARGE.

Table 3. *Observational Behaviors Indicating Pain to Parents*

Observational Behaviors Indicating Pain	***n***
Vocal:	
Crying or tearing up	21
Screaming	3
Moaning or whimpering	2
Social:	
Irritable/cranky/easily upset	4
Less interaction with others (non-sociable) or withdrawn	4
Seeks comfort from parents	3
Pushes other away (e.g. doctors, parents)	2
Obstinate (e.g. doesn't respond to directions, prefers time-out)	1
Acts guarded	1
Facial:	
Grinds teeth	6
Change in eyes (e.g. closes eyes, eyes become glassy)	3
Furrowed brow	1
Crinkled up face	1
Sucks on tongue	1
Activity:	
Lethargic	2
Quieter than normal or change from normal behavior	4
Inconsolable	2
Fussing or not happy	2
Decreased movement	2
Restless/Decreased interest in enjoyable activities (e.g. video games)	2
Body and Limbs:	
Grabs, rubs, or favors part of the body that hurts (e.g. stomach, ear, and chest)	11
	4

Table 3. *Observational Behaviors Indicating Pain to Parents (continued)*

Observational Behaviors Indicating Pain	***n***
Moving body in a specific way to show pain (e.g. curling up, slumped over, and throwing body back)	
Becoming tense or stiff	2
Flexing body parts	1
Retching with any movement or touch	1
Physiological:	
Fever/bruising/pale, flush, or splotchy appearance	4
Days between bowel movements	1
Diaper rash	1
Congestion	1
Holds breath	1
Eating/Sleeping:	
Tired	3
Changes in sleep (e.g. sleepy or refusing to sleep alone)	3
Decreased eating	2
Behavioral Challenges/Self-Injurious and Dangerous Behaviors	
Hits head	4
Acts out/misbehaving	3
Throws/beats up objects (e.g. favorite blanket)	2
Aggressive (e.g. hitting parents or temper tantrums)	2
Bites hand	2
Punches	2
Frustrated	1
Pulls out g-tube	1

Parents also rated their child's pain intensity and average duration of pain (over one year) for common characteristics of CHARGE. A moderate negative correlation between the mean intensity of pain and the mean duration of pain among children with CHARGE was found, ρ =-.34. There was a tendency for the intensity of pain to increase for those sources of pain that were of shorter duration; therefore, more intense pain ratings were found during periods of acute pain than during periods of chronic pain. One limitation to this finding is that no parent or child could reliability identify chronic pain;

therefore, this may suggest that non-vocal pain behaviors are more difficult to detect over extended episodes of pain, perhaps due to accommodation or built up tolerance for pain.

Children with CHARGE experience considerable amounts of pain, as presented in Table 4. The most intense pain for children with CHARGE, as rated by parents, is migraines followed by constipation, surgery pain, chronic result otitis media (ear infections), sinusitis, gastroesophageal reflux, and breathing. Stratton and Hartshorne (2010) also found that children with CHARGE experience considerable chronic pain (experienced for 95 days or more each year) including: gastroesophageal reflux, difficulty swallowing, breathing difficulty, hip/back pain, abdominal migraines, and muscle pain.

Table 4. *Hierarchy of Most Intense Painful Experiences and Average Duration*

	Pain Intensity			**Days per Year in Pain**	
Characteristic	*M*	*SD*	Range	*M*	*SD*
Migraine	2.67	.87	2-4	13.50	13.51
Abdominal Migraine	2.45	1.10	1-4	97.47	128.95
Constipation	2.38	.80	1-4	52.25	58.38
Surgery Pain	2.34	.97	1-4	9.52	9.40
Chronic Recurrent Otitis Media	2.24	.99	0-4	22.88	32.18
Sinusitis	2.17	.82	1-4	35.13	41.51
Gastroesophageal Reflux	2.06	1.14	0-4	169.29	133.70
Breathing	2.00	1.03	1-4	108.67	131.82
Hip/Back Pain	1.86	.95	1-4	98.09	144.14
Muscle Pain	1.82	.87	1-3	95.70	136.07
Coughing	1.61	.80	1-3	66.48	99.42
Jaw Discomfort	1.56	.88	1-3	13.22	11.17
Difficulty Swallowing	1.50	.83	1-4	129.00	154.04

Stratton and Hartshorne (2010) had parents' complete no-pain and pain observations of their child's non-vocal behavior. To determine if there was a significant difference between no-pain and pain ratings, paired samples *t*-tests were performed. On the NCCPC-R, mean no-pain ratings (*M*= 38.93, *SD*= 7.91) and mean pain ratings (*M*=61.44, *SD*=19.18) were found to be significantly different, $t(40) = 8.15$, $p = .00$, CI 95% [16.93, 28.10], $d = 1.3$. Similarly, on the PPP a paired samples *t*-test was conducted to evaluate if parents mean ratings for when the child was experiencing pain (*M*=25.62, *SD*=13.06) were greater than mean no-pain ratings (*M*=10.77, *SD*=8.09) and this was also significant, $t(51) = 9.59$, $p = .00$, CI 95% [11.74, 17.96], $d = 1.3$. The NCCPC-R and PPP are able to discriminate when children with CHARGE are in pain from when children are not in pain.

Next, Stratton and Hartshorne (2010) compared baseline (no-pain) ratings to pain ratings to discriminate which items showed the most difference between the two ratings. Items that had at least a one-point mean difference from NCCPC-R No Pain to NCCPC-R Pain were: not moving/less active/quiet (#14), tears (#25), not cooperating/cranky/irritable/unhappy (#5), crying (#2), moaning/whining/whimpering (#1), less interaction with others/withdrawn (#6), turning down of mouth/not smiling (#11), being difficult to distract/not able to satisfy or pacify (#8), and a furrowed brow (#9). Items with at least a one-point difference on the PPP are very similar to items from the NCCPC-R: cheerful (#1), cries/moans/screams/whimpers (#4), is sociable/responsive (#2), grimaces/screws up face/screws up eyes (#9), frowns/has furrowed brow/looks worried (#10), and hard to console/comfort (#5). These items are likely behaviors many children with CHARGE would display when experiencing discomfort or pain. Table 5

presents in hierarchical order the mean difference score on the NCCPC-R and PPP from no-pain to pain.

Table 5. *Mean Difference Scores from Baseline to Pain in Descending Order on the NCCPC-R and PPP*

NCCPC-R Item (Item #)	Diff. Score	PPP Item (Item #)	Diff. Score
Not moving, less active, quiet (14)	1.49	Is cheerful (1)*	1.67
Tears (25)	1.17	Cries/moans/screams/ whimpers (4)	1.19
Not cooperating, cranky, irritable, unhappy (5)	1.15	Is sociable/responsive (2)*	1.15
Crying (2)	1.15	Grimaces/screws up face/eyes (9)	1.13
Moaning, whining, whimpering (1)	1.12	Frowns/furrowed brow/looks worried (10)	1.12
Less interaction w/ others, withdrawn (6)	1.12	Hard to console/comfort (5)	1.10
Turning down of mouth, not smiling (11)	1.10	Appears withdrawn/depressed (3)	0.92
Being difficult to distract, not able to satisfy or pacify (8)	1.07	Reluctant to eat/difficult to feed (7)	0.83
Furrowed brow (9)	1.00	Restless/agitated/distressed (13)	0.81
Change in eyes, including: squinching of eyes, eyes opened wide, eyes frowning (10)	0.88	Resists being moved (17)	0.79
Protecting, favoring or guarding part of the body that hurts (19)	0.83	Has disturbed sleep (8)	0.73
Eating less, not interested in food (28)	0.83	Looks frightened (11)	0.56
Change in color, pallor (23)	0.80	Tenses/stiffens/spasms (14)	0.52
Sharp intake of breath, gasping (26)	0.80	Tends to touch rub particular areas (16)	0.50
Gesturing to or touching part of the body that hurts (18)	0.76	Twists & turns/tosses head/arches back (19)	0.44
Lips puckering up, tight, pouting, or quivering (12)	0.73	Pulls away or flinches when touched (18)	0.40

Table 5. *Mean Difference Scores from Baseline to Pain in Descending Order on the NCCPC-R and PPP (continued)*

NCCPC-R Item (Item #)	Diff. Score	PPP Item (Item #)	Diff. Score
Moving the body in a specific way to show pain (21)	0.73	Self harms (6)	0.29
Clenching or grinding teeth, chewing or thrusting tongue out (13)	0.71	Grinds teeth or making mouthing movements (12)	0.25
Seeking comfort or physical closeness (7)	0.61	Flexes inward/draws legs up (15)	0.19
Stiff, spastic, tense, rigid (17)	0.61	Involuntary/Stereotypical movements/is jumpy/ startles/seizures (20)	0.15
Increase in sleep (29)	0.61		
Decrease in sleep (30)	0.59		
Flinching or moving the body part away, being sensitive to touch (20)	0.54		
Specific sound, or word for pain (4)	0.51		
Sweating, perspiring (24)	0.51		
Breath holding (27)	0.44		
Screaming/yelling (3)	0.27		
Floppy (16)	0.15		
Shivering (22)	0.15		
Jumping around, agitated, fidgety (15)	0.10		

*Please note higher scores on these items indicate such behaviors are not present or present very little when in pain.

Several behaviors, however, appeared not to display a meaningful difference between pain and no pain across both measures including: flexing inward/drawing legs up (PPP #15), stereotypical movements/jumping/seizures (PPP # 20), flopping (NCCPC-R #16), shivering (NCCPC-R #22), and jumping around/agitation/fidgety (NCCPC-R #

15). Such behaviors may not be useful indicators of pain for use with this population due to their common presentation in CHARGE. As a result, the inclusion of such items will not accurately discriminate pain from no-pain when applied to individuals with CHARGE.

Lastly, Stratton and Hartshorne (2010) asked parents to indicate which instrument, either the NCCPC-R or PPP, best described their child's pain and why. Fifty-five percent of parents (*n*=18) stated the PPP best described their child's pain, whereas 45% (*n*=15) found the NCCPC-R be to more descriptive of pain for their child. Further, more parents (*n*=11) completed the PPP than the NCCPC-R. Based on the results, it appears many parents had difficulty determining which assessment was a better measure of pain for their child, because 20 parents either left this item blank or indicated both measures were helpful. Parental comments are presented in Table 6.

Table 6. *Parental Reasoning for Determining Which Instrument Best Described Their Child's Pain*

NCCPC-R	**PPP**
• "Felt they were similar, but the NCCPC-R allows for more differentiation between the things [my child] can do and the things they can't do."	• Describes more of the characteristics or my child's pain behaviors *(Indicated by 3 parents)*
• "It helped address some of the clues [to my child's pain] better."	• "Goes along with the verbal behavior/communication my child has." *(Indicated by 2 parents)*
• "Allows for you to focus on one area at a time—very easy to follow by heading as things occurred/are observed."	• "A little easier to follow as I went along." • "Easier to complete and score." • "Visually easier to use."

Table 6. *Parental Reasoning for Determining Which Instrument Best Described Their Child's Pain (continued)*

NCCPC-R	PPP
• "I liked how the categories were broken down."	• "Easier to use because my child doesn't experience chronic, all day pain. PPP seemed more suited for [my child]."
• "The categories matched the pain my child experiences better."	• "May relate to infrequent pain episodes better."
• "Body language is a large part of my child's communication."	• "Hard to know when my child is in pain, so if I had to guess, this would probably be better."
• "More specific cues…I was able to see subtle changes that are key [to observe pain]." *(Indicated by 2 parents)*	
• "I believe it is a bit more comprehensive." *(Indicated by 2 parents)*	
• "Allows for better answers and help quicker."	
• "[Both] clearly define what to look for instead of asking to describe pain on a 1-10 scale. My child doesn't understand the 1-10!"	
• "Provides a quantifiable number to compare pain levels on different days or periods of time."	
• "Thought both had good ways to measure pain. Both were descriptive."	
• "The PPP is probably easier and quicker to use, but the NCCPC-R is much more detailed and categorized."	
• "Observation is key to understand [my child's] pain."	
• "I like using both together. I really believe both the NCCPC-R and PPP should be used together to describe symptoms/behaviors with pain, especially when the child cannot communicate very effectively. Each one [scale] reinforces the other's description. It can be very hard to understand what our kids are going through and in turn, difficult to get that across to doctors and therapists when needed."	

Nearly 80% of parents believed the non-vocal, multidimensional pain measurements were helpful in assessing the amount of pain their child was experiencing, whereas 2% indicated the scales were "maybe helpful." Additionally, 83% of parents agreed that the use of such scales would be beneficial in the child's classroom, employment setting, or the like, and 10% indicated they "may be helpful" in these settings. These results demonstrate the need for such instruments not only for parents, but also in clinical, educational, and healthcare settings. Proper identification across settings is helpful for intervention.

Overall, acute pain is a serious concern for individuals with CHARGE and further investigation is needed to determine how chronic pain impacts the lives of individuals with CHARGE. If the pain remains unidentified and untreated, the pain severity and distress the child experiences will increase. Additionally, when communication is limited and parents have difficulty identifying the pain, challenging behaviors are likely to increase as the child attempts to communicate their pain to others. Such challenging behaviors, may be the indicators of pain that are missed by some parents, as evidenced by the 25% of parents who indicated they had difficulty detecting their child's pain experiences.

While the NCCPC-R and PPP were found to be useful instruments to identify pain in children with CHARGE, many items did not seem useful for this population. Additionally, with the exception of one item identifying self-injurious behaviors on the PPP, these scales do not include categories of behavior identified as important by many parents in Stratton and Hartshorne's (2010) study, such as dangerous, aggressive, and self-injurious behaviors. A non-vocal, multidimensional pain assessment tool designed

specifically for CHARGE could help parents and others accurately identify pain. Once the pain is identified, treatment options can be explored and the likelihood of challenging behaviors could be reduced.

Summary

Bottos and Chambers (2006) stress that considerable research needs to be conducted to identify pain among individuals with particular disabilities and the specific types or sources of pain these individuals experience. For individuals with CHARGE syndrome there may be many sources of pain given the nature of their syndrome. However, the actual identification of pain non-vocally can be difficult for this population as the pain induced behavior they display may not be typical for other individuals with developmental disabilities or genetic syndromes. As a result, the following study is designed to investigate a non-vocal pain assessment instrument designed specifically for individuals with CHARGE, the CHARGE Non-Vocal Pain Assessment (CNVPA).

CHAPTER III

RESEARCH QUESTIONS

The goal of this project was to identify non-vocal behavioral signs of pain accurately and objectively for individuals with CHARGE syndrome using the CNVPA. The purpose of this study was to determine the reliability and validity the CNPVA in this frequently non-vocal population. Accurate identification and measurement of pain will also allow for improved clinical management, as well as research into the impact of pain on behavior and learning. The following questions were proposed:

1. Is the CNVPA reliable?

   1a. Does this instrument demonstrate high stability (test-retest)?

   1b. Does this instrument demonstrate high internal consistency?

   1c. Does this instrument demonstrate inter-rater reliability?

2. Does the CNVPA demonstrate validity, indicating support for using this measure for individuals with CHARGE?

   2a. Does the CNVPA demonstrate concurrent validity when correlated to pain ratings on the NCCPC-R and PPP?

   2b. Does the CNVPA display construct validity by discriminating between when children with CHARGE are in pain and when children are not in pain?

   2c. Do non-vocal pain behaviors differ based on the age and/or sex (male and female) of the individual with CHARGE?

CHAPTER IV

METHODOLOGY

Participants

Participants were recruited from the CHARGE Syndrome Foundation and through the CHARGE syndrome Yahoo! Groups Listserv and Facebook page. A mailing was also sent to individuals on the research list for the CHARGE Syndrome Research Laboratory at CMU and to those who participated in a previous study conducted by Stratton and Hartshorne (2010) identifying pain in CHARGE. The reason for multiple recruitment efforts were to not limit the age of the child with CHARGE, given that very little research has been conducted on adult non-vocal pain instruments (Breau & Burkitt, 2009). This was particularly important because limitations in communication and behavior may persist in adulthood and complicate the assessment of pain; further, pain identification in adults with CHARGE has not been previously explored.

Measures

Pain in CHARGE Syndrome Demographics Sheet

The Pain in CHARGE Syndrome Demographics Sheet was designed to be completed by parents/caregivers who have a child with CHARGE syndrome (Appendix C). This form was designed specifically for this study and items included basic demographic information and diagnostic information. Parents/caregivers completed a checklist indicating the presense of major or minor characteristics of CHARGE for their child. Developmental items are also included, e.g., walking age, sleep problems, number of surgeries the child has experienced, and how well the child hears and sees. Further,

this questionnaire captured the child's primary communication strategies, e.g., speech, sign language, communication board/sign, own method, combination, and primarily behavioral/lacking formal language.

CHARGE Non-Vocal Pain Assessment (CNVPA)

The CNVPA was generated through the work of a previous study investigating the identification of pain for children with CHARGE (Stratton & Hartshorne, 2010). Items from the NCCPC-R and PPP with at least a .7 difference score between no-pain and pain observations and additional items, indicated by parents to be useful behavioral identifiers of pain for their child, comprise the CHARGE non-vocal pain assessment instrument. In Stratton and Hartshorne's (2010) investigation, parents indicated that the subscales on the NCCPC-R were helpful to their assessment of their child's pain; therefore, this format was used on the CNVPA. The CNVPA has 30-items divided into five subscales: vocal, social, facial, activity/challenging behaviors, and body and limbs/physiological. Items from the NCCPC-R and PPP were used with permission from the authors for each scale.

Parents also indicated in Stratton and Hartshorne's (2010) study that the Likert-rating on the PPP was easier to complete and score than that used by the NCCPC-R; consequently, this scoring system was used in the creation of this instrument. Parents and caregivers rated their child's behaviors on a 4-point Likert scale: 0-not at all, 1-a little, 2-quite a lot, and 3-a great deal. The ratings were to be completed after a daily observation of the child. The CNVPA can be found in Appendix D.

Procedure

This project was approved at the Institutional Review Board at Central Michigan University. Parents/caregivers were solicited to participate in this study through the CHARGE Syndrome Foundation, the CHARGE Yahoo! Group listserv, the CHARGE Facebook group, and through mailings to previous participants of the CHARGE Syndrome Research Laboratory at CMU. Once parents contacted us to participate (those who were solicited through the CHARGE Syndrome Foundation email, the CHARGE Yahoo! Group listserve, and the CHARGE Facebook page), a mailing was sent to each family with two copies of the consent form (one copy for the parent/caregiver and one copy to return), the pain in CHARGE syndrome demographic form, and the CHARGE non-vocal pain assessment instruments. Materials were sent with a pre-paid return envelope addressed to the CHARGE Syndrome Laboratory at Central Michigan University.

Mailings were sent to parents from research mailing lists at the CMU CHARGE Research Laboratory and the CHARGE Syndrome Foundation using the Dillman Method (2007). The Dillman Method (2007) is a tailored-design for mail surveys that has been shown to increase response rates. First, participants were contacted with a pre-notice letter sent a few days prior to the questionnaire. Second, the questionnaire packet was mailed along with a stamped returned addressed envelope. The questionnaire packet included two copies of the consent form (one for the family to keep and one to return), a cover letter, a demographics form, and the CHARGE non-vocal pain assessments. Third, a thank-you post card was mailed to indicate the packet had been received and a reminder postcard was sent to families twice, if the packet had not been returned. Lastly, a final

thank you will be sent at study completion to highlight the results for the parents/caregivers. If the CNVPA is found to be a reliable and valid instrument for this population, it will also be provided in the final mailing.

Parents/caregivers completed the Pain in CHARGE Demographics sheet first. Second, parents/ caregivers completed the CNVPA during a time when the child was not suspected of being in pain (e.g., watching television or eating breakfast). This served as a baseline measurement of the child's non-vocal behavior. Next, parents/caregivers completed the CNVPA during a time of pain (e.g., migraine headache, abdominal migraine, sinusitis, or injury). Parents were also asked to complete a second pain assessment after a period of no pain since the first pain assessment. The second completion was designed to investigate test-retest reliability. The period of no pain between the two assessments was desired to ensure parents were not rating the same pain episode as the one observed in the first pain assessment. Each questionnaire included the date of the assessment to allow for calculation of the test-retest interval. Parents were encouraged to complete the checklists a short time after the painful event to ensure adequate results, but no later than by the end of the day.

Stratton and Hartshorne's (2010) participants completed two other non-vocal pain assessments, the PPP and NCCPC-R. Participants of Stratton and Hartshorne's study were also be recruited for this study. These participants were asked if their previous data could be used for the present project using the CNVPA. This allowed for an investigation of concurrent validity, by comparing their ratings of pain on the PPP and NCCPC-R with the CHARGE non-vocal pain assessment instrument.

CHAPTER V

RESULTS

Participants

Participants were recruited through a variety of means. Individuals were solicited directly (i.e. Stratton & Hartshorne 2010 participants and the CMU CHARGE syndrome laboratory participants) and through the CHARGE Yahoo! Listserv and Facebook pages. Packets were mailed to 237 families either through the Dillman Method or through a parent's direct request to participate. There were 77 responses, resulting in a 32% response rate, and of these 62 were able to be used. Five were returned for wrong address/unable to forward, four participants sent an email indicating they would not be able to participate (e.g. their child lived in a residential facility or away from the home), two packets were returned with insufficient data completed and two children were deceased. Two remaining packets were returned stating 1) the child living at that address did not have CHARGE and 2) "this packet does not apply to my daughter."

Participant Demographics

Mothers completed 88.5% (*n*= 54) of the packets, fathers completed 11.5% (*n*= 7) of the packets, and one did not specify. Male children comprised 62.3% (*n*=38) of the sample and mean age for the individual with CHARGE was 15.25 years of age (*SD*=8.25 years), with a range from 7 months to 41.5 years of age.

All participants' children had a clinical diagnosis of CHARGE. The average age of diagnosis was 15 months, ranging from less than one month to 20 years of age. A CHARGE diagnosis was mostly made by a geneticist (78.3%, *n*=47), according to parent

report. Clinical diagnosis was also reported to be made by an: ENT (*n*=5), pediatrician (*n*=2), family physician (*n*=1), neonatologist (*n*=1), pre-neonatal physician (*n*=1), ophthalmologist (*n*=1), and two did not specify. In this investigation, 39.3% (*n*=24) of the sample had been genetically tested for the *CHD7* gene mutation, known to produce CHARGE. Of the remaining participants, none were tested and one participant chose to not complete the demographic questionnaire. Of those tested for the *CHD7* mutation, 67% (*n*=16) tested positive. Genetic testing was completed from 1995 to 2011. It should be noted that children tested prior to 2004 were informed several years later, because the *CHD7* gene was only identified in 2004 (Vissers et al., 2004). Table 7 provides the percentages of CHARGE characteristics, or major features of CHARGE, present in this study (*n*= 61).

Table 7. *Frequency of CHARGE Syndrome Characteristics*

CHARGE Syndrome Characteristic/Feature	***n***	**Frequency (%)**
Coloboma of the eye	53	86.9
CHARGE outer ear	51	83.6
Heart defects	47	77.0
Typical CHARGE face	47	77.0
CHARGE inner ear	44	72.1
Vestibular problems	44	72.1
Growth deficiency	43	70.5
CHARGE middle ear	42	68.9
Obsessive-compulsive behavior or perseverative behavior	40	65.6
Choanal atresia or stenosis	38	62.3
Cranial nerve dysfunction/anomaly	35	57.4
Genital abnormalities (hypoplasia)	34	55.7
Kidney/renal abnormalities	28	45.9
Cleft lip +/- cleft palate	18	29.5
Palm crease (hockey-stick palmar crease)	19	31.1
TE (Tracheosophageal) fistula	14	23.0

Children in this study also had significant vision and hearing problems despite corrective measures, e.g., eyeglasses, hearing aids, or other hearing devices. As indicated in Table 8, approximately 60% of the sample displayed a range of vision impairments with correction (e.g., eyeglasses) from moderately impaired to complete blindness. Even more significantly impacted was hearing with approximately 70% of children with CHARGE in this investigation displaying a range between moderate hearing deficits to complete Deafness with correction (e.g., hearing aids, Cochlear implant, etc).

Table 8. *Frequency of Vision and Hearing Deficits* (*n*=61*)*

	Frequency of Vision and Hearing Deficits (%)			
	Vision Left Eye	Vision Right Eye	Hearing Left Ear	Hearing Right Ear
Normal	18.0	16.4	8.2	19.7
Some trouble seeing/hearing	21.3	24.6	18.0	18.0
Moderate difficulty seeing/hearing	21.3	27.9	18.0	19.7
Much difficulty seeing/hearing	27.9	18.0	19.7	11.5
Totally blind/Deaf	11.5	13.1	36.1	31.1

Several other developmental milestones were reported (*N*=61). Fifty-four participants were walking at the time of this study (88.5%). Of those walking, the average age children began walking was 3.5 years of age (*SD*=1.40), with a range of 1.50 years to 7 years of age. Further, at the time of this study, 77% of individuals with CHARGE ate by mouth (*n*=47), 39.3% were tube fed (*n*=24), and 16.4% ate by a combination of mouth and tube (*n*=10). Additionally, 63.9% had problems with sleep (*n*=39) (Table 9). Many individuals had difficulty staying asleep and falling asleep, while fewer had teeth grinding problems and pain during sleep, as noted by parent/caregiver report.

Table 9. *Types and Frequency of Sleep Difficulties*

Sleep Problem	***n***	**Frequency (%)**
Difficulty staying asleep	25	41.0
Difficulty falling asleep	22	36.1
Problems breathing during sleep	12	19.7
Teeth grinding in sleep	12	19.7
Pain during sleep	5	8.2

Parents/caregivers were also asked to indicate their child's communication skills. A nearly bimodal distribution of communication skill was reported (Table 10). Of these individuals with CHARGE, 26.2% (*n*=16) communicated by "making reactions or noises or behaviors which need to be interpreted, and that are difficult for an unfamiliar person to understand." Further, 42.6% (*n*=26) used verbal or sign language in complete sentences. The other participants ranged in communication skill from gestures and body movements to the use of 2 to 5 word phrases or symbols (e.g., sign language, picture symbols).

Table 10. *Parent/caregiver Report of Child's Communication Skill (as adapted from HomeTalk)*

Communication skill	*n*	Frequency (%)
Uses verbal or sign language in complete sentences	26	42.6
Uses some 2 to 5 word phrases and sentences using speech, signs, picture symbols, etc.	7	11.5
Uses single words, signs, pictures, or object symbols to represent basic needs	5	8.2
Uses behaviors such as gestures, sounds, and body movements which most people can interpret or understand	7	11.5
Makes reactions or noises or behaviors which I need to interpret, and that are difficult for an unfamiliar person to understand	26	26.2

Parents were asked to report if their child experienced pain from any of the following painful experiences commonly reported in individuals with CHARGE (Stratton & Hartshorne, 2010): ear infections, sinus infections, gastroesophageal reflux, constipation, surgery pain, tactile defensiveness, migraines, stoma pain, abdominal migraines, muscle pain, back pain, hip pain, and jaw pain. Results are reported in Table 11. In addition, the number of surgeries in this sample (n=56), ranged from 1 to 63 with an average of 13 surgeries (M=12.76, SD=11.77). Ear infections, sinus infections, gastroespophgeal reflux, and constipation were the most frequently reported pain experiences among these participants.

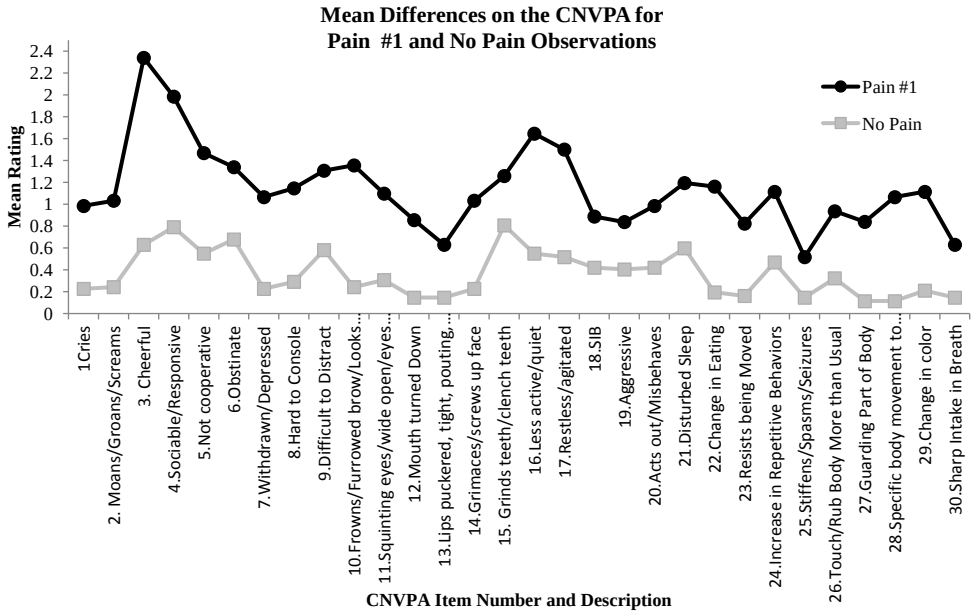
Table 11. *Parent Reported Frequency of Pain Characteristics Commonly Noted for CHARGE* (*N*=61)

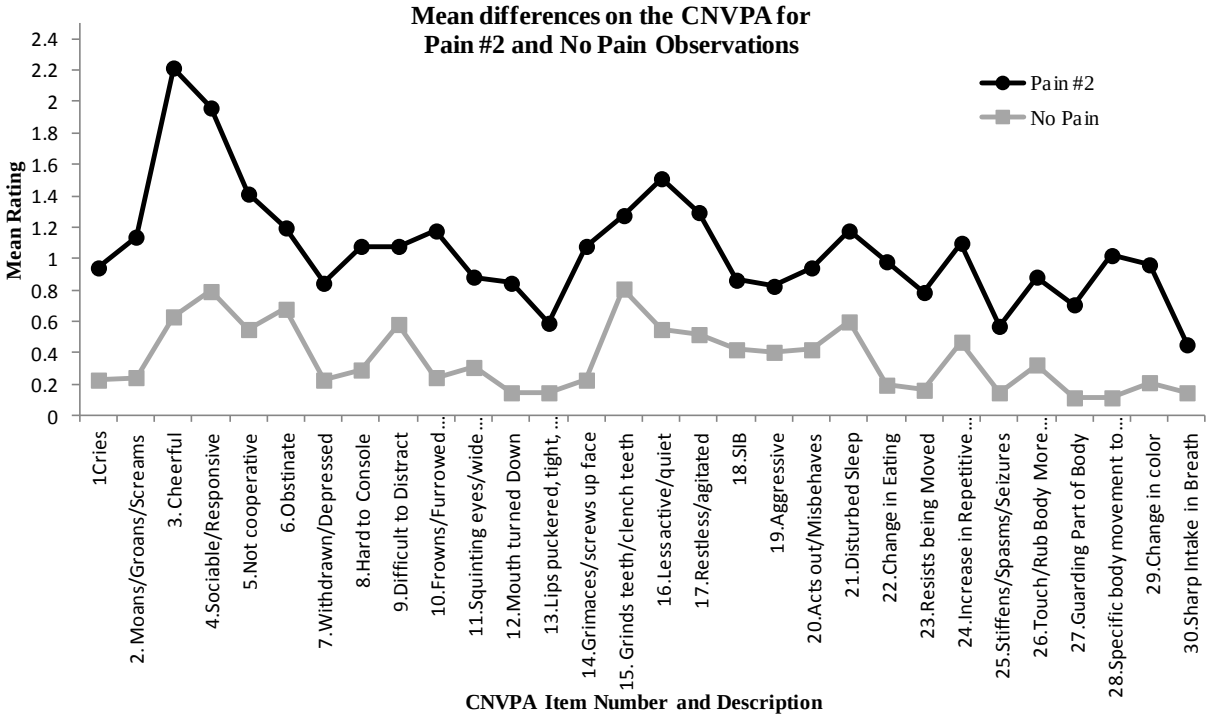
Pain	***n***	**Frequency (%)**
Ear infections	41	67.2
Sinus infections	27	44.3
Gastroesophageal reflux	26	42.6
Constipation	26	42.6
Surgery pain	23	37.7
Tactile defensiveness	21	34.4
Migraine	15	24.6
Stoma pain	12	19.7
Abdominal migraine	12	19.7
Muscle pain	12	19.7
Back pain	8	13.1
Hip pain	6	9.8
Jaw pain	5	8.2

Research Question 1: Is the CNVPA reliable?

An item analysis of the CNVPA is presented in Graph A that includes mean ratings of both no-pain and pain items across both pain ratings (Pain #1 and Pain #2). Baseline ratings (no-pain observations) were compared to pain ratings and several behaviors were shown to have at least a one-point mean difference.

Graph A.


Mean Differences on the CNVPA for Pain #1 and No Pain Observations
Mean Rating
2.4
2.2
2
1.8
1.6
1.4
1.2
1
0.8
0.6
0.4
0.2
0
Pain #1
No Pain
1Cries
2. Moans/Groans/Screams
3. Cheerful
4.Sociable/Responsive
5.Not cooperative
6.Obstinate
7.Withdrawn/Depressed
8.Hard to Console
9.Difficult to Distract
10.Frowns/Furrowed brow/Looks...
11.Squinting eyes/wide open/eyes...
12.Mouth turned Down
13.Lips puckered, tight, pouting...
14.Grimaces/screws up face
15. Grinds teeth/clench teeth
16.Less active/quiet
17.Restless/agitated
18.SIB
19.Aggressive
20.Acts out/Misbehaves
21.Disturbed Sleep
22.Change in Eating
23.Resists being Moved
24.Increase in Repetitive Behaviors
25.Stiffens/Spasms/Seizures
26.Touch/Rub Body More than Usual
27.Guarding Part of Body
28.Specific body movement to...
29.Change in color
30.Sharp Intake in Breath
CNVPA Item Number and Description



*Please note: Items 3 and 4 indicate the child is not cheerful and not sociable/responsive when in pain.

Mean difference scores between pain and no pain (highest difference score to the lowest) are presented in Table 12 for Pain #1 and Pain #2. Items with at least a one-point mean difference from baseline to pain ratings on the CNVPA Pain #1 administration were: not cheerful, aggressive, not sociable/responsive, frowns/furrowed brow/looks worried, and less active/quiet. Items demonstrating nearly a one-point mean difference between baseline and pain ratings on the CNVPA were: restless/agitated, change in eating, specific body movement to indicate pain, not cooperative, and change in color. Similar items were found on a second pain administration. Items with at least a one-point mean difference from baseline to pain ratings on the second administration of the CNVPA were not cheerful and not sociable/responsive. Items with nearly a one-point

mean difference between baseline and pain ratings were: specific body movement to indicate pain, frowns/furrowed brow/looks worried, and less active/quiet. All items had at least a .30 mean difference between baseline and pain ratings, indicating that nearly all items demonstrated a meaningful difference score across no pain and pain experiences.

Table 12. *Mean Difference Scores from Baseline to Pain in Descending Order on the CNVPA for Pain #1 and Pain #2 Administrations*

CNVPA Item (Item #)	**Pain #1 Difference Score (Pain #1 and Baseline N=62)**	**CNVPA Item (Item #)**	**Pain #2 Difference Score (Pain #2 N=51, Baseline N=62)**
*Cheerful (3)	1.70	* Cheerful (3)	1.58
Aggressive (19)	1.24	*Sociable/Responsive (4)	1.17
*Sociable/Responsive (4)	1.19	Less active/quiet (16)	0.96
Frowns/Furrowed brow/Looks Worried (10)	1.11	Frowns/Furrowed brow/Looks Worried (10)	0.93
Less active/quiet (16)	1.09	Specific body movement to indicate pain (28)	0.90
Restless/agitated (17)	0.98	Moans/Groans/Screams (2)	0.89
Change in Eating (22)	0.96	Not cooperative (5)	0.86
Specific body movement to indicate pain (28)	0.95	Grimaces/screws up face (14)	0.85
Not cooperative (5)	0.91	Hard to Console (8)	0.78
Change in color (29)	0.90	Change in Eating (22)	0.78
Hard to Console (8)	0.85	Restless/agitated (17)	0.77

Table 12. *Mean Difference Scores from Baseline to Pain in Descending Order on the CNVPA for Pain #1 and Pain #2 Administrations (continued)*

CNVPA Item (Item #)	**Pain #1 Difference Score (Pain #1 and Baseline N=62)**	**CNVPA Item (Item #)**	**Pain #2 Difference Score (Pain #2 N=51, Baseline N=62)**
Withdrawn/ Depressed (7)	0.83	Change in color (29)	0.75
Grimaces/screws up face (14)	0.80	Cries (1)	0.71
Moans/Groans/ Screams (2)	0.79	Mouth turned Down (12)	0.69
Squinting eyes/wide open/eyes frowning (11)	0.79	Increase in Repetitive Behaviors (24)	0.63
Cries (1)	0.75	Resists being Moved (23)	0.62
Difficult to Distract (9)	0.72	Withdrawn/Depressed (7)	0.61
Guarding Part of Body (27)	0.72	Guarding Part of Body (27)	0.59
Mouth turned Down (12)	0.70	Disturbed Sleep (21)	0.57
Obstinate (6)	0.66	Squinting eyes/wide open/eyes frowning (11)	0.57
Resists being Moved (23)	0.66	Touch/Rub Body More than Usual (26)	0.55
Increase in Repetitive Behaviors (24)	0.64	Acts out/Misbehaves (20)	0.52
Touch/Rub Body More than Usual (26)	0.61	Obstinate (6)	0.51
Disturbed Sleep (21)	0.59	Difficult to Distract (9)	0.49

Table 12. *Mean Difference Scores from Baseline to Pain in Descending Order on the CNVPA for Pain #1 and Pain #2 Administrations (continued)*

CNVPA Item (Item #)	**Pain #1 Difference Score (Pain #1 and Baseline N=62)**	**CNVPA Item (Item #)**	**Pain #2 Difference Score (Pain #2 N=51, Baseline N=62)**
Acts out/Misbehaves (20)	0.48	SIB (18)	0.44
Lips puckered, tight, pouting, quivering (13)	0.48	Lips puckered, tight, pouting, quivering (13)	0.44
Sharp Intake in Breath (30)	0.48	Lips puckered, tight, pouting, quivering (13)	0.44
SIB (18)	0.46	Stiffens/Spasms/ Seizures (25)	0.42
Grinds teeth/clench teeth (15)	0.45	Aggressive (19)	0.42
Stiffens/spasms/seizures (25)	.37	Sharp Intake in Breath (30)	0.30

*Please note higher scores on these items indicate such behaviors are not present or present very little when in pain.

Research Question 1a: Does This Instrument Demonstrate High Stability (test-retest)?

In this investigation, parents completed the CNVPA across two episodes of pain, Pain #1 and Pain #2, to determine the stability of the CNVPA. The dates of each assessment were collected and an average retest interval was 14 days (M=13.82), with a range of 1 to 55 days between Pain #1 and Pain #2 ratings on the CNVPA. Test-retest reliability was examined using Pearson's correlation coefficient and a high stability was found across administrations, r=.77. According to Bracken (1987) and Cicchetti (1994), an appropriate criterion for test-retest reliability is .90. For low-stakes testing or

screening instruments, such as the CNVPA, a stability coefficient of .80 is reasonable (Wasserman & Bracken, 2003).

Table 12 presents the mean differences for both pain ratings (pain #1 and pain #2) from baseline observations. Mean differences are presented in a descending order for all items. Both administrations display very similar mean difference scores; however, the mean differences are slightly lower for pain #2 observations.

Research Question 1b: Does This Instrument Demonstrate High Internal Consistency?

According to Bracken (1987) the internal consistency of an assessment should be .90 or greater. Screening instruments typically require a reliability coefficient of .80 or greater (Wasserman & Bracken, 2003). Cronbach's alpha was used to determine the internal consistency of the CNVPA for both Pain #1 ratings (N=62) and Pain #2 (N=51) ratings. Both ratings demonstrated high internal consistency, $\alpha = .93$ and .94, respectively, according to Bracken's (1987) criterion. Cronbach's alpha was also used to calculate the internal consistency of the CNVPA for the baseline administration, $\alpha = .91$ (N=62).

Research Question 1c: Does This Instrument Demonstrate Inter-rater Reliability?

In an optional portion of this study, parents/caregivers were asked to have another rater (familiar to the child, e.g. spouse, nurse, teacher) complete the CNVPA at the same time as the parent. For pain assessment #1, eight secondary raters completed a pain measurement and five additional raters completed the pain #2 assessment. An intraclass correlation coefficient was used to calculate inter-rater reliability. According to Cicchetti (1994) .75 to 1.00 indicates excellent inter-rater reliability. For the first pain

measurement *r*= .78 and *r*=.98 for the second measurement, providing preliminary evidence for consistency in total scores across raters familiar to the child.

Research Question 2: Does the CNVPA Demonstrate Validity?

Research Question 2a: Does This Instrument Demonstrate Concurrent Validity When Compared to Pain Ratings on the NCCPC-R and PPP?

Given the very limited non-vocal pain assessments available for individuals with CHARGE, the CNVPA was correlated with the NCCPC-R and PPP, based on results from Stratton and Hartshorne's (2010) study. Pearson's correlation was used to examine the relationship of the CNVPA total score (for both Pain #1 and Pain #2) with the total scores of the NCCPC-R and the PPP. CNVPA Pain #1 (N=15) and Pain #2 (N= 13) had similar significant correlations with the NCCPC-R, *r*=.52 and *r*=.65, respectively. Additionally, similar significant correlations were also found between CNVPA Pain #1 (N=20) and #2 (N=15) with the PPP, *r*=.524 and *r*=.516. These results also indicate the CNVPA provide similar yet somewhat different measures of pain for this population from other non-vocal pain measures (NCCPC-R and PPP). It should be noted that the range of time from NCCPC-R and PPP completion and completion of the CNVPA was 14-20 months. Despite the substantial amount of time between completions of the pain assessments they correlated well.

Research Question 2b: Does the CNVPA Display Construct validity by Discriminating between When Individuals with CHARGE Are in Pain and When Individuals Are Not in Pain?

Parents scored the items on the CNVPA at baseline (no pain) and again when their child was in pain. The expectation was that there would be a considerable

difference in the ratings on the items for the two administrations because ratings at baseline should be lower than ratings during pain, as the CNVPA was developed to measure pain. As such, higher ratings would occur during pain. The CNVPA was also found to be a stable instrument with adequate test-retest reliability (r=.77). Therefore, to evaluate whether parents' ratings for pain were greater than ratings for when the child was suspected to not have pain, a paired samples t-test was conducted.

On the first pain administration (Pain #1) of the CNVPA the mean rating on the items when the child was in pain was 34.13 (SD=17.23) and the ratings for when they were not in pain was 10.85 (SD=9.56). The difference between these ratings was significant, t (61) = 11.17, p<.001, CI 95% [19.17, 27.44]. The effect size was d =1.4, a large effect.

A second CNVPA pain administration was completed by parents (Pain #2). The mean rating on the second administration when the child was in pain was 31.76 (SD=17.27) and the ratings for when they were not in pain was 11.27 (SD=9.72). The baseline (no pain) mean reported is based on a smaller sample size for the second administration of Pain #2 (N=51). Similar to the Pain #1 and baseline ratings, the difference between Pain #2 and no pain ratings were significant, t (50) = 8.31, p<.001, CI 95% [15.54, 25.44]. The effect size was d =1.2, a large effect. On both administrations, all of the items were rated lower when the child was not in pain than when the child was in pain.

Parents were also asked to provide what type of pain their child experienced during the time the CNVPA was completed. Across both pain ratings, common painful experiences described were: ear and sinus infections, constipation, headaches/migraines,

surgery, G-tube complications/gastroesphageaul reflux, and falls and/or accidents. Other less frequent pain experiences included: leg, back, hip, and neck pain, pneumonia, hernia, spinal infection, bone fracture, and adjustment of braces. Additionally, parents were asked to report if the pain experience identified during the administration of the CNVPA was confirmed. Confirmation was provided using the following scale (see Table 13): child vocally told me, physiological symptoms (e.g. fever, cough, runny nose), bruise, the behavior displayed was common when the child experienced that type of pain, and physician confirmed pain (e.g. ear infection, sinus infection, broken bone) with an open ended option for diagnosis. Parents provided confirmation of pain on both administrations of the CNVPA. On the first administration, approximately 38% of individuals with CHARGE were able to vocally express their pain experience, providing confirmations that the parents' observations on the CNVPA are of pain experiences. Additionally, 36% of our sample had a confirmed diagnosis known to produce pain by a physician, further displaying construct validity of the CNVPA. Similar results were found during the second pain administration as displayed in Table 14.

Table 13. *Parent Reported Confirmation of Child's Pain: Pain 1 (Note: Parents were told to check all that apply). (N=61)*

Confirmation of Pain	***n***	**Frequency (%)**
Behavior displayed is common when my child experiences this type of pain	42	68.9
Child vocally told me	23	37.7
Physician confirmed pain (e.g. ear/sinus infection, broken bone)	22	36.1
Physiological symptoms (e.g. fever, cough, runny nose)	13	21.3
Bruise	5	8.2

Table 14. *Parent Reported Confirmation of Child's Pain: Pain 2 (Note: Parents were told to check all that apply). (N=50)*

Confirmation of Pain	***n***	**Frequency (%)**
Behavior displayed is common when my child experiences this type of pain	29	58.0
Child vocally told me	18	36.0
Physician confirmed pain (e.g. ear/sinus infection, broken bone)	15	30.0
Physiological symptoms (e.g. fever, cough, runny nose)	10	20.0
Bruise	6	12.0

Research Question 2c: Do Non-vocal Pain Behaviors Differ Based on the Age or Sex of the Individual with CHARGE?

Based on participant responses, six age cohorts were formed: 1) birth to age 5 years 12 months, 2) ages 6 years to 10 years 12 months, 3) ages 11 years to 15 years, 12 months, 4) ages 16 years to 20 years 12 months, 5) ages 21 years to 25 years 12 months, and 6) ages 26 and older. Mean differences between Pain #1 and No Pain ratings on the CNVPA are presented in Table 15 by age cohort and in Graphs B separately by two age cohorts.

It was expected that children ages one month to five years of age would demonstrate fewer, if any, challenging behaviors compared to other age cohorts. Children in this age cohort showed minor differences or no difference between no pain and pain ratings on the CNVPA for the following items (CNVPA item number in parentheses): aggressive (19), acts out/misbehaves (20), guards a part of the body (27), obstinate (6), touch/rub part of the body more than usual (26), self-injurious behaviors (18), and increase in repetitive behaviors (24). Individuals in the other age cohorts generally were rated to have more variability in scores across challenging behavior items on the CNVPA.

Variability by age was noted across multiple age groups. For example, as presented in Graph B, individuals in the oldest age cohort (26 years of age and older) had nearly a one point or more mean difference score (between no pain and pain) higher than all other age cohorts. Other greater difference scores from other age cohorts included: squinting eyes/eyes wide open/eyes frowning (11), mouth turned down (12), self-injurious behaviors (18), disturbed sleep (21), resists being moved (23), and a specific body movement to indicate pain (28). The youngest age cohort (birth to age five), was

rated to have higher mean difference scores than all other age groups on the following three items: change in eating habits (22), less active/quiet (16), and a change in color (29). Parents and caregivers typically report looking for similar changes in behavior for children of this age group. Teenagers in the age cohort of 11 to 15 years of age, were rated as having a lower mean difference score on aggressive behaviors (item 19) and grinding teeth/clenching teeth (item 15) than all other age cohorts.

Across all age cohorts one of the largest mean differences between no pain and pain ratings was on item #3 on the CNVPA: less cheerful. Similarly, item #4, less sociable/responsive, was also rated to be one of the largest mean difference scores between no pain and pain.

Table 15. Mean Differences of CNVPA Items by Age Cohorts (No Pain and Pain #1)

CNVPA Item (Item #)	Age Birth to 5 Years (N=8)	Age 6 to 10 Years (N=14)	Age 11 to 15 Years (N=10)	Age 16 to 20 Years (N=13)	Age 21 to 25 Years (N=10)	Age 26 and Up (N=6)
Cries	1.00	1.00	.50	.69	.50	1.00
Moans/Groans/Screams	1.00	.93	.50	.54	.60	1.50
Cheerful*	1.75	1.93	1.50	1.85	1.30	2.00
Sociable/Responsive*	1.38	1.36	.90	1.62	.50	1.33
Not Cooperative	.75	1.14	1.10	.85	.50	1.17
Obstinate	.25	.71	.70	.62	.60	1.33
Withdrawn or Depressed	.50	.71	.90	1.08	.60	1.50
Hard to Console or Comfort	.75	1.21	.40	.85	.60	1.33
Difficult to Distract	.63	.79	.60	.85	.20	1.67
Frowns/Has Furrowed Brow/Looks Worried	1.38	1.00	.50	1.54	.40	2.33

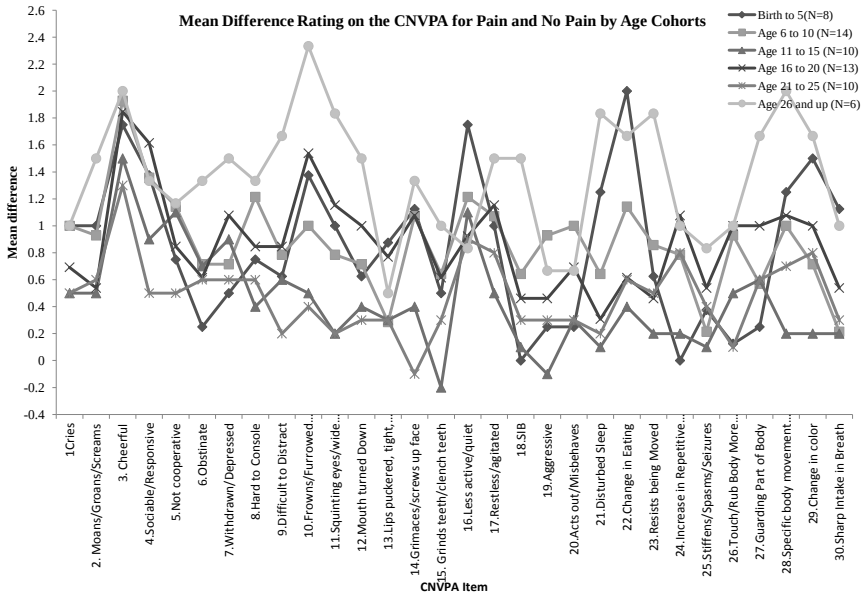
Table 15. Mean Differences of CNVPA Items by Age Cohorts (No Pain and Pain #1) (continued)

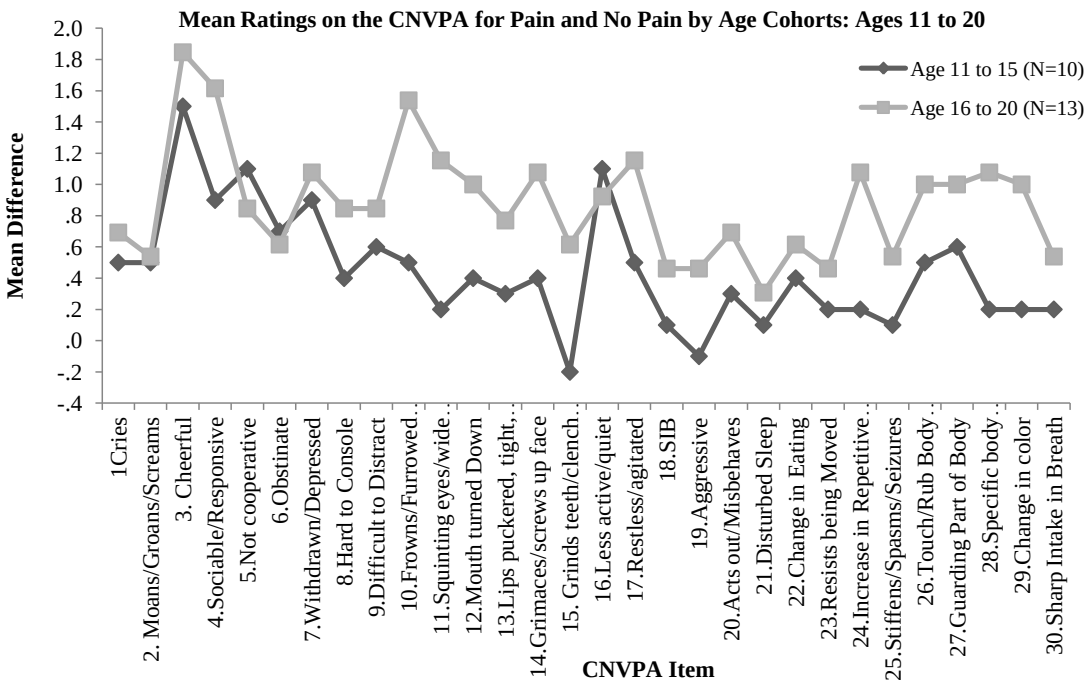
CNVPA Item (Item #)	Age Birth to 5 Years (N=8)	Age 6 to 10 Years (N=14)	Age 11 to 15 Years (N=10)	Age 16 to 20 Years (N=13)	Age 21 to 25 Years (N=10)	Age 26 and Up (N=6)
Squinting Eyes/Eyes Wide Open/Eyes Frowning	1.00	.79	.20	1.15	.20	1.83
Mouth Turned Down	.63	.71	.40	1.00	.30	1.50
Lips Puckered Up, Tight, Pouting, or Quivering	.88	.29	.30	.77	.30	.50
Grimaces/Screws up Face	1.13	1.07	.40	1.08	-.10	1.33
Grinds Teeth/Clenches Teeth	.50	.64	-.20	.62	.30	1.00
Less Active or Quiet	1.75	1.21	1.10	.92	.90	.83
Restless/Agitated	1.00	1.07	.50	1.15	.80	1.50
Self-Injurious Behaviors	0.00	.64	.10	.46	.30	1.50
Aggressive	.25	.92	-.10	.46	.30	.67
Acts out/Misbehaves	.25	1.00	.30	.69	.30	.67

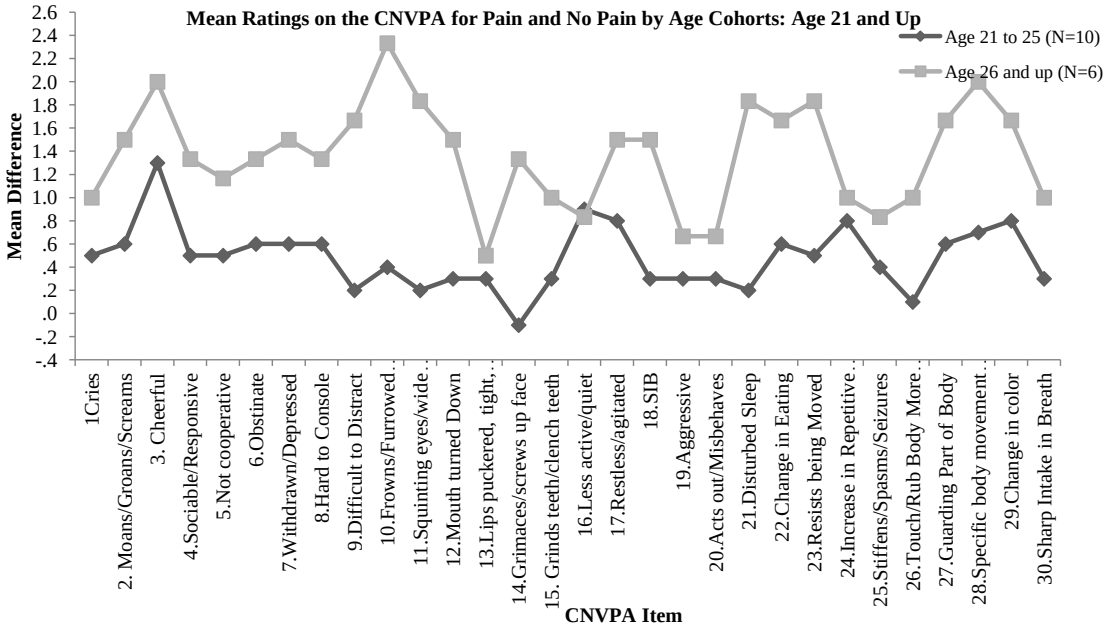
Table 15. Mean Differences of CNVPA Items by Age Cohorts (No Pain and Pain #1)(continued)

CNVPA Item (Item #)	Age Birth to 5 Years (N=8)	Age 6 to 10 Years (N=14)	Age 11 to 15 Years (N=10)	Age 16 to 20 Years (N=13)	Age 21 to 25 Years (N=10)	Age 26 and Up (N=6)
Disturbed Sleep	1.25	.64	.10	.31	.20	1.83
Change in Eating Habits	2.00	1.14	.40	.62	.60	1.67
Resists Being Moved	.63	.86	.20	.46	.50	1.83
Increase in Repetitive Behaviors	0.00	.79	.20	1.08	.80	1.00
Stiffens/Spasms/Seizures	.38	.21	.10	.54	.40	.83
Touching or Rubbing Parts of the Body More than Usual	.13	.93	.50	1.00	.10	1.00
Guarding a Part of the Body	.25	.57	.60	1.00	.60	1.67
Specific Body Movement to Indicate Pain	1.26	1.00	.20	1.08	.70	2.00
Change in Color	1.5	.71	.20	1.00	.80	1.67
Sharp Intake of Breath/Gasping	1.13	.21	.20	.54	.30	1.00
Stiffens/Spasms/Seizures	.38	.21	.10	.54	.40	.83
Touching or Rubbing Parts of the Body More than Usual	.13	.93	.50	1.00	.10	1.00

Graph B.


Mean Difference Rating on the CNVPA for Pain and No Pain by Age Cohorts
Birth to 5(N=8)
Age 6 to 10 (N=14)
Age 11 to 15 (N=10)
Age 16 to 20 (N=13)
Age 21 to 25 (N=10)
Age 26 and up (N=6)
Mean difference
2.6
2.4
2.2
2
1.8
1.6
1.4
1.2
1
0.8
0.6
0.4
0.2
0
-0.2
-0.4
1Cries
2. Moans/Groans/Screams
3. Cheerful
4.Sociable/Responsive
5.Not cooperative
6.Obstinate
7.Withdrawn/Depressed
8.Hard to Console
9.Difficult to Distract
10.Frowns/Furrowed...
11.Squinting eyes/wide...
12.Mouth turned Down
13.Lips puckered, tight...
14.Grimaces/screws up face
15. Grinds teeth/clench teeth
16.Less active/quiet
17.Restless/agitated
18.SIB
19.Aggressive
20.Acts out/Misbehaves
21.Disturbed Sleep
22.Change in Eating
23. Resists being Moved
24. Increase in Repetitive...
25.Stiffens/Spasms/Seizures
26.Touch/Rub Body More...
27.Guarding Part of Body
28.Specific body movement...
29.Change in color
30.Sharp Intake in Breath
CNVPA Item


Mean Ratings on the CNVPA for Pain and No Pain by Age Cohorts: Ages 11 to 20
Age 11 to 15 (N=10)
Age 16 to 20 (N=13)
Mean Difference
2.0
1.8
1.6
1.4
1.2
1.0
.8
.6
.4
.2
.0
-.2
-.4
1Cries
2. Moans/Groans/Screams
3. Cheerful
4.Sociable/Responsive
5.Not cooperative
6.Obstinate
7.Withdrawn/Depressed
8.Hard to Console
9.Difficult to Distract
10.Frowns/Furrowed..
11.Squinting eyes/wide..
12.Mouth turned Down
13.Lips puckered, tight,..
14.Grimaces/screws up face
15. Grinds teeth/clench..
16.Less active/quiet
17.Restless/agitated
18.SIB
19.Aggressive
20.Acts out/Misbehaves
21.Disturbed Sleep
22.Change in Eating
23.Resists being Moved
24.Increase in Repetitive..
25.Stiffens/Spasms/Seizures
26.Touch/Rub Body..
27.Guarding Part of Body
28.Specific body..
29.Change in color
30.Sharp Intake in Breath
CNVPA Item


Mean Ratings on the CNVPA for Pain and No Pain by Age Cohorts: Age 21 and Up
Age 21 to 25 (N=10)
Age 26 and up (N=6)
Mean Difference
2.6
2.4
2.2
2.0
1.8
1.6
1.4
1.2
1.0
.8
.6
.4
.2
.0
-.2
-.4
1Cries
2. Moans/Groans/Screams
3. Cheerful
4.Sociable/Responsive
5.Not cooperative
6.Obstinate
7.Withdrawn/Depressed
8.Hard to Console
9.Difficult to Distract
10.Frowns/Furrowed..
11.Squinting eyes/wide..
12.Mouth turned Down
13.Lips puckered, tight,..
14.Grimaces/screws up face
15. Grinds teeth/clench teeth
16.Less active/quiet
17.Restless/agitated
18.SIB
19.Aggressive
20.Acts out/Misbehaves
21.Disturbed Sleep
22.Change in Eating
23.Resists being Moved
24.Increase in Repetitive..
25.Stiffens/Spasms/Seizures
26.Touch/Rub Body More..
27.Guarding Part of Body
28.Specific body movement..
29.Change in color
30.Sharp Intake in Breath
CNVPA Item

Sex differences were also explored on the CNVPA. CNVPA items were rank ordered for each sex (male or female) to explore differences. Table 16 and Graph C present the mean differences between pain #1 and no pain across sex on the CNVPA.

Table 16. *Mean Differences of CNVPA Items by Sex in Descending Order (No Pain and Pain #1)*

CNVPA Item (Item #)	**Mean Difference Male (N=38)**	**CNVPA Item (Item #)**	**Mean Difference Female (N=23)**
*Cheerful (3)	1.68	*Cheerful (3)	1.78
Less active/quiet (16)	1.26	*Sociable/Responsive (4)	1.30
Frowns/Furrowed brow/Looks Worried (10)	1.18	Withdrawn/Depressed (7)	1.13
*Sociable/Responsive (4)	1.13	Change in color (29)	1.08
Specific body movement to indicate pain (28)	1.13	Frowns/Furrowed brow/Looks Worried (10)	1.00
Not cooperative (5)	1.05	Change in Eating (22)	1.00
Restless/agitated (17)	1.02	Restless/agitated (17)	.91
Change in Eating (22)	.97	Moans/Groans/Screams (2)	.91
Hard to Console (8)	.92	Less active/quiet (16)	.86
Squinting eyes/wide open/eyes frowning (11)	.86	Cries (1)	.73
Grimaces/screws up face (14)	.86	Hard to Console (8)	.73
Mouth turned Down (12)	.81	Difficult to Distract (9)	.73

Table 16. *Mean Differences of CNVPA Items by Sex in Descending Order (No Pain and Pain #1) (continued)*

CNVPA Item (Item #)	**Mean Difference Male (N=38)**	**CNVPA Item (Item #)**	**Mean Difference Female (N=23)**
Cries (1)	.78	Increase in Repetitive Behaviors (24)	.73
Resists being Moved (23)	.78	Guarding Part of Body (27)	.73
Change in color (29)	.78	Not cooperative (5)	.69
Acts out/Misbehaves (20)	.76	Grimaces/screws up face (14)	.69
Difficult to Distract (9)	.73	Squinting eyes/wide open/eyes frowning (11)	.69
Guarding Part of Body (27)	.73	Disturbed Sleep (21)	.69
Moans/Groans/Screams (2)	.71	Touch/Rub Body More than Usual (26)	.69
Obstinate (6)	.71	Specific body movement to indicate pain (28)	.69
Withdrawn/Depressed (7)	.68	Obstinate (6)	.60
Increase in Repetitive Behaviors (24)	.63	Mouth turned Down (12)	.56
Aggressive (19)	.60	Lips puckered, tight, pouting, quivering (13)	.52
Touch/Rub Body More than Usual (26)	.60	Resists being Moved (23)	.47
SIB (18)	.55	Stiffens/Spasms/Seizures (25)	.43

Table 16. *Mean Differences of CNVPA Items by Sex in Descending Order (No Pain and Pain #1) (continued)*

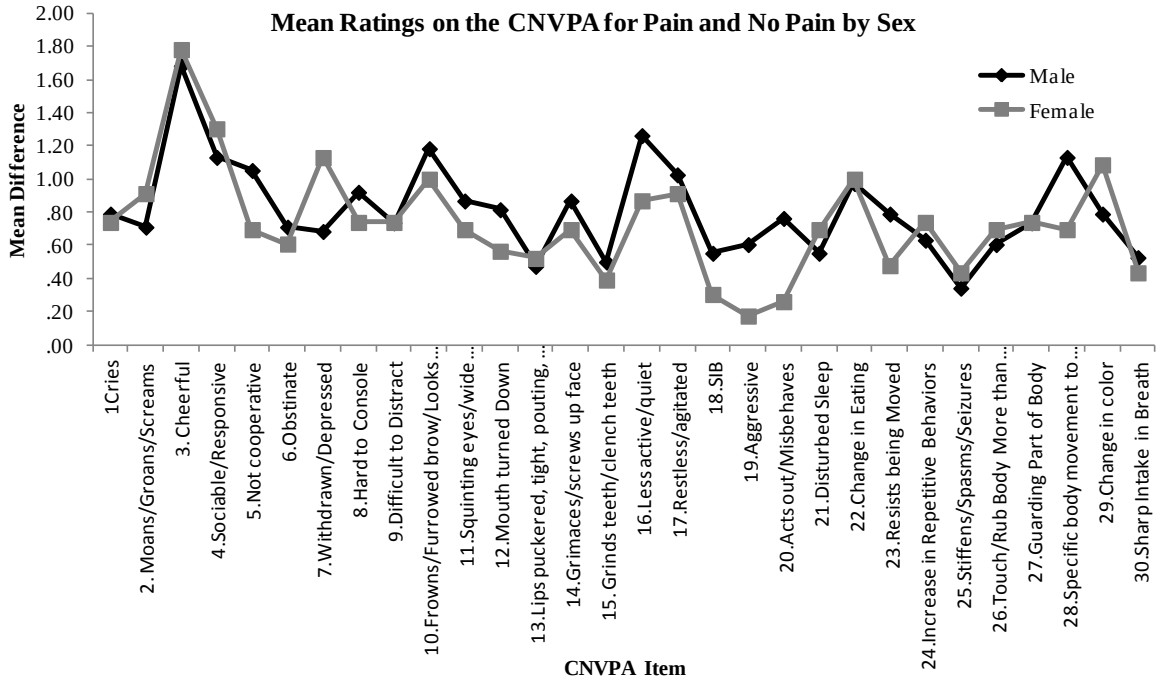
CNVPA Item (Item #)	**Mean Difference Male (N=38)**	**CNVPA Item (Item #)**	**Mean Difference Female (N=23)**
Disturbed Sleep (21)	.55	Sharp Intake in Breath (30)	.43
Sharp Intake in Breath (30)	.52	Grinds teeth/clench teeth (15)	.39
Grinds teeth/clench teeth (15)	.50	SIB (18)	.30
Lips puckered, tight, pouting, quivering (13)	.47	Acts out/Misbehaves (20)	.26
Stiffens/Spasms/Seizures (25)	.34	Aggressive (19)	.17

* Please note higher scores on these items indicate such behaviors are not present or present very little when in pain.

Individuals with CHARGE, regardless of their sex, typically presented with similar mean difference scores on the CNVPA. While similar mean difference ratings were found, some items demonstrated a difference between male and female individuals with CHARGE. Females had a higher mean difference score on item #7 (withdrawn or depressed) and item #29 (change in color). Males were rated with higher mean difference scores on facial items (item number in parentheses): frowns/has furrowed brow/looks worried (10), squinting eyes/eyes wide open/eyes frowning (11), and mouth turned down (12). Males were also rated as having higher mean difference scores on not cooperative (5) less active/quiet (16), aggression (19), acts out/misbehaves (20), and specific body movement to indicate pain (28). Generally, fewer differences in mean

difference scores between CNVPA no pain and pain ratings were found when compared to a variety of differences between age cohorts.

Graph C.



Research Question 3d: Does This Instrument Display Face Validity or Acceptance by the Parents/Caregivers?

Face validity or social acceptability of the CNVPA was assessed by asking parents if they found the CNVPA to be relevant to identify pain (non-vocally) for their child on the following Likert-rating scale (Item #3; Appendix D): Not relevant, somewhat relevant, relevant, or extremely relevant. Based on the results presented in Table 17, approximately 85% of parents and secondary observers endorsed the CNVPA to be a relevant ("somewhat relevant" to "extremely relevant") assessment to identify pain for individuals with CHARGE.

Table 17. *Parental (N=59) and Secondary Observer (N=8) Report of CNVPA Relevance to Identifying Child's Pain*

	Parental Response		**Secondary Observer Response**	
	n	**Frequency (%)**	***n***	**Frequency (%)**
Not Relevant	9	15.3	1	12.5
Somewhat Relevant	18	30.5	2	25.0
Relevant	18	30.5	5	62.5
Extremely Relevant	14	23.7	0	0

Additionally, parents were asked to provide why they did not find the CNVPA to be relevant. Eight parents responded to this item and indicated a variety of reasons why the CNVPA was not relevant to identifying pain for their child. For four participants, the scale was rated as "not relevant" because the child had vocal communication and could state when they were experiencing pain. Two parents indicated that they had already developed behavioral cues for their child when they were experiencing pain (e.g., "We already know pain indicators." "We know when he acts out that pain is present."). Lastly, one parent indicated that even with the CNVPA "it is difficult to discern if something is wrong." Another responded the scale was not relevant "because when [my child] is sick, he is actually easier to take care of… he just wants to be left alone laying down and occasionally whimpering." One secondary observer noted that the CNVPA was not relevant to identify pain in the child observed because they could vocalize their pain experience.

Many parents (*n*=13) and three secondary observers chose to use the open-ended option on the CNVPA Item 3 (Why?) to provide information on why they found the scale to be relevant, although this sub-question was intended for parents who did not find the

scale to be relevant. Descriptive responses for parents (and secondary observers) that endorsed the CNVPA to be relevant are presented in Table 18.

Table 18. *Descriptive Explanations of Why Parents (and secondary observers) Endorsed the CNVPA to be a Relevant Assessment to Identify their Child's Pain.*

Relevance of CNVPA to Identify Pain (Likert relevance rating provided in parentheses with the following range: somewhat relevant, relevant, extremely relevant)
Vocalizations: • "She is verbal, but will rarely complain of pain." (Relevant) • "Child is verbal." (Somewhat Relevant) • "Child is vocal." (Somewhat Relevant) • "Child is vocal and will tell people to leave her alone." (Relevant) • "Has full verbal communication, but he does become more quiet when an ear infection is beginning." (Somewhat Relevant) • "My child never complains of pain and seems to tolerate it very well. He is not usually a complainer." (Relevant)
Items describe or help identify when my child is in pain: • "Observations and questions match exactly how my child reacts." (Extremely Relevant) • "After 24 years, I'm very in tune with my child's health and items match." (Somewhat Relevant) • "I've already developed ways to identify changes in my child's behavior." (Somewhat Relevant) • "My child doesn't show much outwardly when in pain, other than verbalizing it and bad behavior." (Somewhat Relevant) • "This information confirms our experience in assessing our son's pain through the years." (Relevant) • "She is variable." (Somewhat Relevant) • "It is hard to know why she is irritated or more agitated, we usually don't know if there is pain until we see a bruise or a fever." (Somewhat Relevant) • "Helpful for me to compare her behaviors when ill to behaviors when she is well. There is a big distinction between the two." (Relevant) • "We have to notice emotional behavior for this child." (Secondary observer; Relevant) • "Easy to tell by behaviors." (Secondary observer; Relevant)

CHAPTER VI

DISCUSSION

This investigation is an extension of Stratton and Hartshorne's (2010) first research study of pain among individuals with CHARGE. Both studies found that individuals with CHARGE experience many painful episodes throughout their lives, both acute and chronic (Stratton & Hartshorne, 2010). In the current study, parents pointed to multiple surgical procedures in addition to pain from ear infections, sinus infections, gastroesophageal reflux, and constipation as primary pain experiences.

There are several complicating factors to understanding pain experiences for this population. First, many individuals with CHARGE are unable to vocally communicate or use formal language (verbal or signed) to indicate their pain experiences and seek treatment. Second, significant hearing and visual impairments likely impact the ability to communicate effectively to others the subjective experiences of pain. In addition, hearing and visual impairments further delay children with CHARGE from learning socially appropriate ways to react to pain from peer modeling. Stratton and Hartshorne (2010) found that individuals with CHARGE appear to display some different non-vocal pain behaviors than individuals with other developmental delays, based on results of two non-vocal pain measurements (NCCPC-R and the PPP). As a result, the development and validation of a CHARGE specific non-vocal pain assessment seemed necessary.

The present study provides preliminary evidence that when used in a natural setting the CNVPA is a reliable and valid measure when used by parents and other significant caregivers to indicate the presence of pain. The analyses presented provide evidence for moderate to strong psychometric properties.

The first finding from this study is that the CNVPA approached acceptable levels for reliability. Test-retest reliability is a measure of temporal stability. One of the challenges of rating pain episodes is that it is unknown how stable pain is over time, particularly given the subjective nature of pain. For example, Hunt et al. (2004) found that for thirty individuals with developmental delay and severe neurological disability, average pain scores on the PPP did not differ across postoperative surgery ratings for up to five days after surgery (gastrointestional or orthopedic surgery). The highest pain score, however, was found within the first 24 hours following surgery for nearly half of the sample. This variability in pain stability may influence test-retest results. In addition, it is unknown for individuals with CHARGE if protective pain behaviors (e.g. decreased activity) and pain severity influence temporal stability. Additional data are needed to determine the stability of different types of pain for individuals with CHARGE and how this may influence test-retest reliability and the need for treatment.

A second and important analysis of reliability is internal consistency. This is an indicator of uniformity among the assessment items. Findings in the present study are similar to the NCCPC-R (Breau et al., 2002). Hunt et al.'s (2004) investigation of reliability indicated the PPP had an internal consistency of .83 and .86 for two separate pain ratings, somewhat lower than the CNVPA. This finding is important because it shows that the CNVPA has strong internal consistency similar to other established non-vocal pain assessments. Further, this finding indicates that the CNVPA has a strong consistency of scores when rated by parents within a naturalistic environment (e.g. home).

Multiple raters were found to have strong agreement when rating pain non-vocally using the CNVPA. This finding is important because it provides preliminary evidence that items on the CNVPA are well enough described that different raters tend to observe the same behavior. It should be noted that this study had a very small sample and future studies are needed to further explore inter-rater agreement of the CNVPA. However, the interrater coefficients from this study were consistent with findings for two previously established non-vocal pain instruments, the PPP and NCCPC-R (Breau et al., 2002; Hunt et al. 2004).

The value of the CNVPA was first established by its strong reliability; however, the second set of analyses addressed to the validity of the CNVPA. There was evidence that the CNVPA has good concurrent validity, when compared to other established non-vocal pain assessment instruments, the PPP and NCCPC-R, given to individuals with CHARGE. Cicchetti (1994) stated that it is not possible to create a "rule of thumb" for concurrent validity because such decisions rely on what the new assessment purports to measure in relation to the previously established assessments. The CNVPA is designed to measure pain for individuals who CHARGE, whereas previously established measures have been designed for populations of individuals with developmental delay or neurological disabilities. It was expected that the CNVPA would have a moderate correlation (.40 to .60) when compared to the previously established measurements (PPP and NCCPC-R) considering that the CNVPA measures similar behaviors to those on the PPP and NCCPC-R but included additional behaviors identified by parents of individuals with CHARGE to indicate pain.

Another important finding from this study is evidence to support construct validity. The finding that the CNVPA clearly discriminated when individuals were in pain from when they were not in pain is noteworthy because the CNVPA is a tool that can be used to assist parents who are having difficulty making the distinction. Baseline ratings (e.g. completion of the CNVPA on a "good day" or when pain is not suspected) can serve as markers for parents and can be used for comparison with ratings when pain is suspected. Professionals working with the family (e.g. school psychologists, behavioral psychologists, teachers) might assist parents who want to develop baseline and pain ratings using the CNPVA and graph data over time to better understand their child's pain experiences.

In addition, confirmation of the pain experience provides further evidence of construct validity. In addition to higher ratings on the CNVPA from baseline during pain assessment, many individuals in this study were able to vocalize their pain experience or physicians' confirmed pain. This indicates that parental ratings of suspected pain behaviors using the CNVPA are appropriate and meaningful, particularly for their children who do not have formal communication (vocal or sign).

The CNVPA was also found to yield similar pain ratings for both sexes (male and female), indicating the CNVPA is equally applicable to measure pain non-vocally for both males and females. Based on the results from this study, males tended to score higher on challenging behavior items. This may indicate that the challenging behaviors described by parents of children with CHARGE are more likely in males than in females. Future studies of the CNVPA will need to further explore the sex differences noted in this investigation.

In this study, the CNVPA did not differ dramatically across age cohorts; however, the youngest and oldest cohorts displayed the largest difference. The oldest age cohort (age 26 and older) exhibited higher mean difference scores from baseline to pain in facial characteristics and challenging behaviors. While additional studies will need to replicate these results, this result suggests that adults with CHARGE typically display challenging behaviors and changes in facial expression when in pain. For adults with limited formal communication (sign or vocal), challenging behaviors may bring about the most attention from caregivers who could provide more immediate attention to the cause of the pain. Additionally, adults with CHARGE may display facial expressions of pain more clearly than younger children because they have learned the social appropriateness of expressing emotions, whereas younger individuals with CHARGE may still have a socio-communicative deficit.

The youngest cohort in this study was rated to have no or very little challenging behavior and demonstrated pain behaviors typical to children in infancy or toddlerhood. This indicates that the pain behaviors of infants and toddlers with CHARGE are easier to identify than pain behaviors displayed by children as they age and become adults. As a result, the CNVPA may prove to be more valuable for individuals with CHARGE and their caregivers after the age of 5 to identify pain non-vocally. All individuals in this investigation, however, displayed a high mean difference score from baseline to pain on two items: not cheerful and not sociable/responsive. These two behaviors can serve as indicators to parents and other significant caregivers that the child may be in pain or experiencing discomfort. Additionally, these behaviors may prompt parents/caregivers the use of the CNVPA to assess pain and determine if medical attention is needed.

Lastly, while the CNVPA was found to have good psychometric properties, an assessment is only relevant if users find the assessment to be useful and to provide information they may not otherwise have access to. Gregory (2011) describes face validity as social acceptability. The CNVPA must look valid to test users. Parents rated the CNVPA as a relevant assessment to identify pain non-vocally. Based on parental responses, however, the CNVPA is likely to be more relevant for individuals with limited or no formal communication to indicate pain, than for individuals with established formal communication (e.g. vocal or signed language) and understanding of subjective experiences like pain. Future studies evaluating the CNVPA would benefit from including additional studies on face validity to further evaluate social acceptability of the CNPVA.

Limitations

This investigation has several limitations. First, the sample size was small despite flooding the market for participants and the return rate was low (36%). There are several possible reasons for the low return rate. First, the packet may have been overwhelming to parents. Second, parents have been repeatedly told their child has a high pain threshold; therefore, parents may have accepted this hypothesis and chosen not to respond to the pain study. Third, parents may not have participated because their child did not experience a clear painful event during the collection period. Further, as Stratton and Hartshorne (2010) have reported, many children with CHARGE experience chronic pain. If the child experiences chronic pain, the parents may have had difficulty completing a baseline assessment, limiting their willingness to participate. Lastly, parents of children who have vocal communication may have not participated in the

study because the results are not relevant to their child or parents may have believed the child's communication would confound results for a non-vocal measure of pain.

Another limitation to this investigation is that it relied on parents and secondary observers familiar to the child to accurately complete the study. While attempts were made to achieve interrater agreement, it is unknown from this investigation whether others, particularly individuals who are not familiar with the child (e.g., nurses, physicians, new care staff), would observe and rate the child's behaviors in the same fashion as parents and others who know the child well.

Although parents indicated their child's pain experience when completing the CNVPA, and for nearly half of the sample the pain experience was confirmed by a physician, parents may not have always been accurate in indicating that the child was in pain. Future studies may investigate if psychometric properties vary based on various pain experiences i.e. surgery pain and pain from ear infections.

Future Directions

Future areas of research include further examination of the validity of the CNVPA. Although this investigation attempted to control for the experience of pain with confirmation of the pain experience (e.g. physician providing a diagnosis of a painful condition) versus rating other behaviors (e.g. stress, unhappiness), further evidence of validity could be evaluated through more controlled investigations, such as completing the CNVPA before and after a surgical procedure. Surgical procedures would be a useful approach to further analyze the validity of the CNVPA because many individuals with CHARGE endure multiple surgical procedures. With proper identification of pain, early

treatment can be provided and the potential for challenging and dangerous behaviors (e.g. self-injurious behaviors) can be reduced.

This study provided preliminary evidence that the age of the child may impact which specific items are the most useful to identify pain non-vocally. This result could be addressed in a future investigation looking specifically at items for a larger sample to determine if the number of items could be reduced to ease ratings based on specific age groupings and to improve psychometric properties by age. Additionally, males may demonstrate more challenging behaviors than females when in pain, according to this investigation. A larger investigation could include a factor analysis that would assist researchers in determining specific sex differences based on the subcategories of the CNVPA.

Additionally, Stratton and Hartshorne (2010) found that individuals with CHARGE experience chronic and acute pain. It is unknown if non-vocal pain behaviors, like those measured on the CNVPA, would change based on the duration of pain experienced. Further, Stratton and Hartshorne (2010) reported that some areas of adaptive functioning were reduced during periods of pain. It would be useful to investigate how activities of daily living and adaptive functioning are inhibited based on the duration of pain experienced. Communication is an important area of adaptive functioning that could be further limited by pain. This relationship needs to be explored for individuals with CHARGE, including how a reduction in communication and increase in pain or increase in the duration of pain are related.

In conclusion, the CNVPA was found to be reliable and valid measure to identify pain non-vocally for individuals with CHARGE syndrome. This study also warrants

future research to further establish the psychometric properties of the CNVPA and to specifically explore psychometric properties across age groups, sex, pain experiences, and multiple raters (i.e. physicians and nurses).

APPENDICES

APPENDIX A

NON-COMMUNITICATING CHILDREN'S PAIN CHECKLIST-REVISED

Non-communicating Children's Pain Checklist – Revised (NCCPC-R)

Child's Name: _____ Birth date: _____ Sex: Male Female

Date Completed: _____ (month/day/year)

Observer: _____

START Time: _____AM/PM STOP Time: _____AM/PM

How much of the day were you able to observe your child: _____ hours

DIRECTIONS:

How often has your child shown these behaviors today? Please circle a number for each item; see guide below. If an item does not apply to your child (for example, your child does not eat solid food or cannot reach with his/her hands), then indicate "not applicable" for that item.

A guide for deciding the frequency of items is below:

0 = **Not present at all** during observation period (Note if the item is not present because the child is not capable of performing that act, it should be scored as "NA"

1 = **Seen or heard rarely** (hardly at all), but is present

2 = **Seen or heard a number of times**, but not continuous (not all the time)

3 = **Seen or heard often, almost continuous** (almost all the time); anyone would easily notice this if they saw your child for a few moments during the observation time

NA = **Not applicable**; child is not capable of performing this action.

I. Vocal

1. Moaning, whining, whimpering (fairly soft)…… ……. 0 1 2 3 NA

2. Crying (moderately loud)…….....…….... 0 1 2 3 NA

3. Screaming/yelling (very loud)…… ……… 0 1 2 3 NA

4. A specific sound or word for pain (e.g., a word, cry or type of laugh)…..... 0 1 2 3 NA

II. Social

5. Not cooperating, cranky, irritable, unhappy…… ……0 1 2 3 NA

6. Less interaction with others, withdrawn…… ....……..... 0 1 2 3 NA

7. Seeking comfort or physical closeness ……..... ... 0 1 2 3 NA

8. Being difficult to distract, not able to satisfy or pacify…….....…. 0 1 2 3 NA

III. Facial

9. A furrowed brow……..…….....…..... 0 1 2 3 NA

10. A change in eyes, including: squinching of eyes, eyes opened wide, eyes frowning…….....……. 0 1 2 3 NA

11. Turning down of mouth, not smiling…… .....…… 0 1 2 3 NA

12. Lips puckering up, tight, pouting, or quivering…… .....……. 0 1 2 3 NA

13. Clenching or grinding teeth, chewing or thrusting tongue out …… .....…... 0 1 2 3 NA

IV. Activity

14. Not moving, less active, quiet…….. …….…….....……..…. 0 1 2 3 NA

15. Jumping around, agitated, fidgety…….....……... 0 1 2 3 NA

V. Body and Limbs

16. Floppy ……. .....…… 0 1 2 3 NA

17. Stiff, spastic, tense, rigid …….....……..… 0 1 2 3 NA

18. Gesturing to or touching part of the body that hurts ……..…….. 0 1 2 3 NA

19. Protecting, favoring or guarding part of the body that hurts ……....…..…. 0 1 2 3 NA

20. Flinching or moving the body part away, being sensitive to touch ....…..... 0 1 2 3 NA

21. Moving the body in a specific way to show pain

(E.g. head back, arms down, curls up, etc.) ……..…… ....……..… 0 1 2 3 NA

VI. Physiological

22. Shivering …….....…..…… 0 1 2 3 NA

23. Change in color, pallor ……...…..…..... 0 1 2 3 NA

24. Sweating, perspiring …… .....….…..…. 0 1 2 3 NA

25. Tears…….....….…… 0 1 2 3 NA

26. Sharp intake of breath, gasping…….....…….. 0 1 2 3 NA

27. Breath holding…….....…… 0 1 2 3 NA

VII. Eating/Sleeping

28. Eating less, not interested in food…… .....……..…….. 0 1 2 3 NA

29. Increase in sleep…….…….....…..…… 0 1 2 3 NA

30. Decrease in sleep……. ……..... ..... 0 1 2 3 NA

STOP Time: _____ AM/PM

To be completed by researcher

SCORE SUMMARY:

I

II

III

IV

V

VI

VII

TOTAL

APPENDIX B

PAEDIATRIC PAIN PROFILE

Paediatric Pain Profile

DIRECTIONS:

For each item please circle the number that best describes your child's behavior when they ***are experiencing pain***.

	Not at all	**A little**	**Quite a lot**	**A great deal**	**Score**
1. Is cheerful	3	2	1	0	
2. Is sociable or responsive	3	2	1	0	
3. Appears withdrawn or depressed	0	1	2	3	
4. Cries/moans/groans/screams or whimpers	0	1	2	3	
5. Is hard to console or comfort	0	1	2	3	
6. Self-harms e.g., biting self or banging head	0	1	2	3	
7. Is reluctant to eat/difficult to feed	0	1	2	3	
8. Has disturbed sleep	0	1	2	3	
9. Grimaces/screws up face/screws up	0	1	2	3	

eyes					
10. Frowns/has furrowed brow/looks worried	0	1	2	3	
11. Looks frightened (with eyes wide open)	0	1	2	3	
12. Grinds teeth or makes mouthing movements	0	1	2	3	
13. Is restless/agitated or distressed	0	1	2	3	
	Not at all	**A little**	**Quite a lot**	**A great deal**	**Score**
14. Tenses/stiffens or spasms	0	1	2	3	
15. Flexes inwards or draws legs up towards chest	0	1	2	3	
16. Tends to touch or rub particular areas	0	1	2	3	
17. Resists being moved	0	1	2	3	
18. Pulls away or flinches when touched	0	1	2	3	
19. Twists and turns/tosses head/writhes or arches back	0	1	2	3	

20. Has involuntary or stereotypical movements/is jumpy/startles or has seizures	0	1	2	3	
Please list other (e.g., laughing): _____					
Please list other: _____					
				TOTAL:	

How often does your child have pain free days? *(Please Circle)*

ALL THE TIME MOST OF THE TIME SOME OF THE TIME HARDLY EVER

Do you think your child has pain even on a good day? *(Please Circle)*

NO PAIN MILD PAIN MODERATE PAIN SEVERE PAIN VERY SEVERE PAIN

When you child is experiencing their most troublesome pain, what word best describes the severity of this pain? *(Please Circle)*

NONE MILD MODERATE SEVERE VERY SEVERE

APPENDIX C

CHARGE SYNDROME DEMOGRAPHICS SHEET

CHARGE Syndrome

Demographics Sheet **Subject #**

---

1. Are you the child's:

   - ❑ MOTHER
   - ❑ FATHER
   - ❑ GUARDIAN
   - ❑ OTHER: _____

2. Child's Date of Birth **(Month/Day/Year)**:_____/_____/_____
3. Child's Gender:

   - ❑ MALE
   - ❑ FEMALE

4. At what age was your child diagnosed as having CHARGE?

   - ❑ BEFORE AGE 1 ____ MONTHS
   - ❑ AFTER AGE 1 ____ YEARS OLD

5. Who made the diagnosis of CHARGE?

- ❑ GENETICIST
- ❑ FAMILY PHYSICIAN
- ❑ NEONATOLOGIST
- ❑ ENT
- ❑ OTHER:_____

6. Has your child been tested for the *CHD7* gene mutation?

   - ❑ NO (Skip to item 7)
   - ❑ YES

     Did your child test positive or negative for the mutation?

     - ❑ POSITIVE
     - ❑ NEGATIVE

     To your best recollection, when was your child tested?

     (Month/Year) _____/_____

7. CHARGE Characteristics your child displays: **(Check all that apply)**

- ❑ Coloboma of the eye
- ❑ Choanal atresia or stenosis
- ❑ Cranial Nerve Dysfunction/Anomaly
- ❑ CHARGE outer ear
- ❑ CHARGE middle ear
- ❑ CHARGE inner ear
- ❑ Vestibular Problems

- ❑ Heart Defects
- ❑ Cleft lip +/- cleft palate
- ❑ TE (Tracheosophageal) fistula
- ❑ Kidney/Renal Abnormalities
- ❑ Genital Abnormalities (Hypoplasia)
- ❑ Growth deficiency
- ❑ Typical CHARGE Face
- ❑ Palm crease (hockey-stick palmar crease)
- ❑ Obsessive-Compulsive Behavior or Perseverative Behavior

8. Check all of the following that produce pain for your child? **(Check all that apply)**

- ❑ Migraine
- ❑ Abdominal Migraine
- ❑ Jaw Pain
- ❑ Surgery Pain
- ❑ Ear Infections
- ❑ Sinus Infections (sinusitis)
- ❑ Gastroesophageal Reflux
- ❑ Constipation
- ❑ Hip Pain
- ❑ Back Pain
- ❑ Muscle Pain
- ❑ Stoma Pain

- ❑ Tactile Defensiveness
- ❑ Other:_____

9. How many surgeries has your child had? _____ Surgeries

10. How does your child eat? **(Check all that apply)**

- ❑ EATS BY MOUTH
- ❑ TUBE FEEDING

11. Is your child walking?

- ❑ NO (Skip to item 12)
- ❑ YES → My child walked at _____years of age

12. Does your child have problems with sleep?

- ❑ NO (Skip to item 13)
- ❑ YES

What sleep problems does your child display?

- ❑ DIFFICULTY FALLING ASLEEP
- ❑ DIFFICULTY STAYING ASLEEP
- ❑ SLEEP BREATHING PROBLEMS
- ❑ TEETH GRINDING IN SLEEP

- ❑ PAIN DURING SLEEP
- ❑ OTHER: _____

13. How well does your child see? **(with glasses or contact lenses, if used)**

LEFT	RIGHT	
❑	❑	NORMAL VISION
❑	❑	SOME TROUBLE SEEING
❑	❑	MODERATE DIFFICULTY
❑	❑	MUCH DIFFICULTY
❑	❑	TOTALLY BLIND

14. How well does your child hear? **(with hearing aids or other hearing devices, if used)**

LEFT	RIGHT	
❑	❑	NORMAL HEARING
❑	❑	SOME TROUBLE HEARING
❑	❑	MODERATE DIFFICULTY
❑	❑	MUCH DIFFICULTY

❑ ❑ TOTALLY DEAF

15. My child's communication skills are best described as: (adapted from HomeTalk)

❑ Makes reactions or noises or behaviors which I need to interpret, and that are difficult for an unfamiliar person to understand.

❑ Uses behaviors such as gestures, sounds, and body movements which most people can interpret or understand.

❑ Uses single words, signs, pictures, symbols, or object symbols to represents basic needs.

❑ Uses some 2 to 5 word phrases and sentences using speech, signs, picture symbols, etc.

❑ Uses verbal or sign language in complete sentences.

APPENDIX D

CHARGE NON-VOCAL PAIN ASSESSMENT (CNVPA)

CNVPA: NO PAIN Assessment

TODAY'S DATE: _____

DIRECTIONS:

Please complete the following rating after observations of your child for one day when you do not think your child experienced pain. For each item, circle the number that best describes your child's behavior when they are not in pain.

If your child does not engage in a behavior when not in pain OR is not capable of performing an action, score this item as *"not at all."*

	Not at all	**A little**	**Quite a lot**	**A great deal**
VOCAL				
Cries	0	1	2	3
Moans/groans/screams	0	1	2	3
SOCIAL				
Cheerful	3	2	1	0
Sociable/responsive	3	2	1	0
Not cooperative (cranky, irritable)	0	1	2	3
Obstinate (e.g. doesn't respond to directions)	0	1	2	3
Withdrawn or depressed	0	1	2	3
Hard to console or comfort	0	1	2	3
Difficult to distract	0	1	2	3
FACIAL				
Frowns/has furrowed brow/looks worried	0	1	2	3

Squinting eyes/eyes wide open/eyes frowning	0	1	2	3
Mouth turned down	0	1	2	3
Lips puckered up, tight, pouting, or quivering	0	1	2	3
Grimaces/screws up face	0 **Not at all**	1 **A little**	2 **Quite a lot**	3 **A great deal**
Grinds teeth/clenches teeth	0	1	2	3
ACTIVITY/CHALLENGING BEHAVIORS				
Less active or quiet	0	1	2	3
Restless/agitated	0	1	2	3
Self-injurious behaviors (Biting self, banging/hitting head)	0	1	2	3
Aggressive (e.g. hitting others, throwing objects)	0	1	2	3
Acts out/Misbehaves	0	1	2	3
Disturbed sleep	0	1	2	3
Change in eating habits	0	1	2	3
Resists being moved	0	1	2	3
Increase in OCD-like behaviors	0	1	2	3
BODY AND LIMBS/PHYSIOLOGICAL				

Stiffens/spasms/seizures	0	1	2	3
Touching or rubbing parts of the body more than usual	0	1	2	3
Guarding a part of the body	0	1	2	3
Specific body movement to indicate pain (e.g. arms down, curled up, head down)	0	1	2	3
Change in color (e.g., pale, splotchy, flush)	0	1	2	3
Sharp intake of breath/gasping	0	1	2	3

CNVPA: #1 PAIN Assessment

TODAY'S DATE: _____

DIRECTIONS:

Please complete the following rating after observations of your child for one day when you believe your child ***was experiencing pain***. For each item, circle the number that best describes your child's behavior during the pain episode.

If your child does not engage in a behavior when in pain OR is not capable of performing an action, score this item as *"not at all."*

	Not at all	**A little**	**Quite a lot**	**A great deal**
VOCAL				
Cries	0	1	2	3
Moans/groans/screams	0	1	2	3
SOCIAL				
Cheerful	3	2	1	0
Sociable/responsive	3	2	1	0
Not cooperative (cranky, irritable)	0	1	2	3
Obstinate (e.g. doesn't respond to directions)	0	1	2	3
Withdrawn or depressed	0	1	2	3

Hard to console or comfort	0	1	2	3
Difficult to distract	0	1	2	3
FACIAL				
Frowns/has furrowed brow/looks worried	0	1	2	3
Squinting eyes/eyes wide open/eyes frowning	0	1	2	3
Mouth turned down	0	1	2	3
Lips puckered up, tight, pouting, or quivering	0	1	2	3
Grimaces/screws up face	0 **Not at all**	1 **A little**	2 **Quite a lot**	3 **A great deal**
Grinds teeth/clenches teeth	0	1	2	3
ACTIVITY/CHALLENGING BEHAVIORS				
Less active or quiet	0	1	2	3
Restless/agitated	0	1	2	3
Self-injurious behaviors (Biting self, banging/hitting head)	0	1	2	3
Aggressive (e.g. hitting others, throwing objects)	0	1	2	3
Acts out/Misbehaves	0	1	2	3

Disturbed sleep	0	1	2	3
Change in eating habits	0	1	2	3
Resists being moved	0	1	2	3
Increase in OCD-like behaviors	0	1	2	3
BODY AND LIMBS/PHYSIOLOGICAL				
Stiffens/spasms/seizures	0	1	2	3
Touching or rubbing parts of the body more than usual	0	1	2	3
Guarding a part of the body	0	1	2	3
Specific body movement to indicate pain (e.g. arms down, curled up, head down)	0	1	2	3
Change in color (e.g., pale, splotchy, flush)	0	1	2	3
Sharp intake of breath/gasping	0	1	2	3

(Move ahead to the next page)

1. **What do you suspect was the cause of your child's pain today? (e.g., ear infection, fall, injury)**

   _____ is the cause of my child's pain today

2. **Was there confirmation your child was in pain? (Check all that apply)**

   - ❑ Child vocally told me
   - ❑ Physiological symptoms (e.g. fever, cough, runny nose)
   - ❑ Bruise
   - ❑ The behavior displayed is common when my child experiences this type of pain
   - ❑ Physician confirmed pain (e.g., ear infection, sinus infection, broken bone)

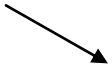
     Diagnosis:

     _____

3. **Did you find the pain assessment relevant to identify pain (non-vocally) for your child?**

   - ❑ EXTREMELY RELEVANT
   - ❑ RELEVANT

- ❑ SOMEWHAT RELEVANT
- ❑ NOT RELEVANT



WHY?

_____

_____

_____

_____

4. **Outside of those behaviors listed on the assessment, does your child display additional behaviors that indicate pain? Please list.**

5. **Please return to the observation items and place a check mark (✓) beside the item that best indicated pain for your child.**

CNVPA: *#2* PAIN Assessment

TODAY'S DATE: _____

DIRECTIONS:

Please complete this assessment during a <u>second period of pain</u>. Your child should have had a period of no pain between the first assessment and this assessment. After a daily observation of your child, circle the number that best describes your child's behavior during the current (2^{nd}) pain episode.

	Not at all	**A little**	**Quite a lot**	**A great deal**
VOCAL				
Cries	0	1	2	3
Moans/groans/screams	0	1	2	3
SOCIAL				
Cheerful	3	2	1	0
Sociable/responsive	3	2	1	0
Not cooperative (cranky, irritable)	0	1	2	3
Obstinate (e.g. doesn't respond to directions)	0	1	2	3
Withdrawn or depressed	0	1	2	3
Hard to console or comfort	0	1	2	3

Difficult to distract	0	1	2	3
FACIAL				
Frowns/has furrowed brow/looks worried	0	1	2	3
Squinting eyes/eyes wide open/eyes frowning	0	1	2	3
Mouth turned down	0	1	2	3
Lips puckered up, tight, pouting, or quivering	0	1	2	3
Grimaces/screws up face	0 **Not at all**	1 **A little**	2 **Quite a lot**	3 **A great deal**
Grinds teeth/clenches teeth	0	1	2	3
ACTIVITY/CHALLENGING BEHAVIORS				
Less active or quiet	0	1	2	3
Restless/agitated	0	1	2	3
Self-injurious behaviors (Biting self, banging/hitting head)	0	1	2	3
Aggressive (e.g. hitting others, throwing objects)	0	1	2	3
Acts out/Misbehaves	0	1	2	3
Disturbed sleep	0	1	2	3

Change in eating habits	0	1	2	3
Resists being moved	0	1	2	3
Increase in OCD-like behaviors	0	1	2	3
BODY AND LIMBS/PHYSIOLOGICAL				
Stiffens/spasms/seizures	0	1	2	3
Touching or rubbing parts of the body more than usual	0	1	2	3
Guarding a part of the body	0	1	2	3
Specific body movement to indicate pain (e.g. arms down, curled up, head down)	0	1	2	3
Change in color (e.g., pale, splotchy, flush)	0	1	2	3
Sharp intake of breath/gasping	0	1	2	3

(Move ahead to the next page)

1. **What do you suspect was the cause of your child's pain today? (e.g., ear infection, fall, injury)**

   _____ is the cause of my child's pain today

2. **Was there confirmation your child was in pain? (Check all that apply)**

   - ❑ Child vocally told me
   - ❑ Physiological symptoms (e.g. fever, cough, runny nose)
   - ❑ Bruise
   - ❑ The behavior displayed is common when my child experiences this type of pain
   - ❑ Physician confirmed pain (e.g., ear infection, sinus infection, broken bone)

   ↘

   Diagnosis:

   _____

3. **Please return to the observation items and place a check mark (✓) beside the item that best indicated pain for your child.**

REFERENCES

American Psychiatric Association. (2004). Diagnostic and statistical manual of mental disorders (4th ed.). Washington, DC: Author.

Blake, K., Davenport, S.H., Hall, B.D., Hefner, M.A., Pagon, R., Williams, M.S.Graham, I.M. (1998). CHARGE association- An update and review for the primary pediatrician. *Clinical Perspectives, 31,* 159-174.

Blake, K.D., Hartshorne, T.S., Lawand, C., Dailor, A.N., & Thelin, J.W. (2008). Cranial nerve manifestations in CHARGE syndrome. *American Journal of Medical Genetics, 146(A),* 585-592.

Blake, K.D., Salem-Hartshorne, N., Daoud, M.A., & Gradstein, J. (2005). Adolescent and adult issues in CHARGE syndrome. *Clinical pediatrics, 44,* 151-159.

Bodfish, J.W., Harper, V.N., Deacon, J.R., & Symons, F.J. (2001, July). Identifying and measuring pain in persons with developmental disabilities: A manual for the pain and discomfort scale (PADS). *Western Carolina Center Research Reports.*

Bottos, S., & Chambers, C.T. (2006). The epidemiology of pain in developmental disabilities. In T.F. Oberlander & F.J. Symons (Eds.), *Pain in children and adults with developmental disabilities* (pp. 67-87). Baltimore, Maryland: Brookes.

Bracken, B.A. (1987). Limitations of preschool instruments and standards for minimal levels of technical adequacy. *Journal of Psychoeducational Assessment, 4,* 313-326.

Breau, L., McGrath, P.J., Finley, A., & Camfield, C. (2004). *Non-communicating children's pain checklist-revised (NCCPC-R).* Halifax, Nova Scotia: Lynn Breau.

Breau, L.M. & Burkitt, C. (2009). Assessing pain in children with intellectual disabilities. *Pain Research, and Management, 14,* 116-120.

Breau, L.M., Camfield, C.S., McGrath, P.J., & Finley, A. (2003). The incidence of pain in children with severe cognitive impairments. *Archives of Pediatric and Adolescent Medicine, 157,* 1219-1226.

Breau, L.M., Camfield, C.S., McGrath, P.J., & Finley, G.A. (2007). Pain's impact on adaptive functioning. *Journal of Intellectual Disability Research,* 51, 125-134.

Breau, L.M., Camfield, C., McGrath, P.J., Rosmus, C., & Finley, G.A. (2001). Measuring pain accurately in children with cognitive impairments: Refinement of a caregiver scale. *The Journal of Pediatrics, 138,* 721-727.

Breau, L.M., McGrath, P.J., Camfield, C.S., & 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., & Zabalia, M. (2006). Assessing pediatric pain and developmental disabilities. In T.F. Oberlander & F.J. Symons (Eds.), *Pain in children and adults with developmental disabilities* (pp. 149-172). Baltimore, Maryland: Brookes.

Brown, D. (2005). CHARGE syndrome "behaviors": Challenges or adaptations? *American Journal of Medical Genetics, 133A,* 268-272.

Cicchetti, D.V. (1994). Guidelines, criteria, and rules of thumb for evaluating normed and standardized assessment instruments in psychology. *Psychological Assessment, 6,* 284-290.

Cook, A.K., Niven, C.A., & Downs, M.G. (1999). Assessing the pain of people with cognitive impairment. *International journal of geriatric psychiatry,* 14, 421-425.

Craig, K.D. (2006). The construct and definition of pain in developmental disability. In T.F. Oberlander & F.J. Symons (Eds.), *Pain in children and adults with developmental disabilities* (pp.7-18). Baltimore, Maryland: Brookes.

Davenport, S.J.H., Hefner, M.A., & Mitchell, J.A. (1986). The spectrum of clinical features in CHARGE syndrome. *Clinical Genetics, 29,* 298-310.

Davenport, S.H. (2002). Overview of CHARGE syndrome for physicians. In M. Hefner & S.H. Davenport (Eds.), *CHAGE syndrome: A management manual for parents 2.1* (pp. 51-52). Columbia, Missouri: CHARGE Syndrome Foundation.

Defrin, R., Lotan, M., & Pick, C.G. (2006). The evaluation of acute pain in individuals with cognitive impairment: A differential effect of the level of impairment. *Pain, 124,* 312-320.

Dillman, D. (2007). Mail and internet surveys: A tailored design method method-Second Edition. Wiley and Sons.

Doobelsteyn, C., Peacocke, S.D., Blake, K., Crist, W., & Rashid, M. (2008). Feeding difficulties in children with CHARGE syndrome: prevalence, risk factors, and prognosis. *Dysphagia, 23,* 127-135.

Ekman, P., & Friesen, W. (1978). *Investigators guide to the facial action coding system.* Palto Alto, CA: Consulting Psychologist Press

Engel, J.M., & Kartin, D. (2006). Pain in individuals with cerebral palsy. In T.F. Oberlander & F.J. Symons (Eds.), *Pain in children and adults with developmental disabilities* (pp. 109-119). Baltimore, Maryland: Brookes.

Gilbert-MacLeod, C.A., Craig, K.D., Rocha, E.M., & Mathias, M.D. (2000). Everyday pain responses in children with and without developmental delays. *Journal of Pediatric Psychology,* 25, 301-308.

Gregory, R. J. (2011). Psychological testing: History, principles, and applications (6th ed.). Boston, MA: Pearson Education, Inc.

Hargreaves, (2007). New migraine and pain research. *Headache, 47S,* 26-43.

Hartshorne, T.S., & Cypher, A.D. (2004). Challenging behavior in CHARGE syndrome. *Mental Health Aspects of Developmental Disabilities, 7*(2), 41-52.

Hartshorne, T.S., Hefner, M.A., & Davenport, S.L.H. (2005). Behavior in CHARGE syndrome. *American Journal of Medical Genetics,* 133A, 228-231.

Hartshorne, T.S., Hefner, M.A., Davenport, S.L.H., Thelin, J. (2010). *CHARGE syndrome.* Plural Publishing.

Hunt, A. (2003). *Paediatric Pain Profile.* Oxford, UK: RCN Institute.

Hunt, A., Goldman, A, Seers, K., Crichton, N., Mastroyannopoulou, K., Moffat, V., Brady, M. (2004). Clinical validation of the paediatric pain profile. *Developmental Medicine & Child Neurology, 46,* 9-19.

Hunt, A., Wisbeach, A., Seers, K., Goldman, A., Crichton, N., Perry, L., Mastroyannopoulou, K. (2007). Development of the paediatric pain profile: role of video analysis and saliva cortisol in validating a tool to assess pain in children with severe neurological disability. *Journal of Pain and Symptom Management, 33,* 276-289.

Issekutz, K.A., Graham, J.M., Prasad, C., Smith, I.M., & Blake, K.D. (2005). An epidemiological analysis of CHARGE syndrome: Preliminary results from a Canadian study. *American Journal of Medical Genetics, 133A,* 309-317.

Iwata, B.A., Pace, G.M., Kissel, R.C., Nau, P.A., & Farber, J.M. (1990). The self-injury trauma (SIT) scale: A method for quantifying surface tissue damage caused by self-injurious behavior. *Journal of Applied Behavior Analysis, 23,* 99-110.

Kangas, M., Henry, J.L., & Bryant, R.A. (2002). Posttraumatic stress disorder following cancer: A conceptual and empirical review. *Clinical Psychology Review, 22,* 499-524.

Kassam-Adams, N. (2006). Introduction to the special issue: Posttraumatic stress related to pediatric illness and injury. *Journal of Pediatric Psychology,* 31, 337-342.

Kennedy, C.H., & O'Reilly, M.F. (2006). Pain, health conditions, and problem behavior in people with developmental disabilities. In T.F. Oberlander & F.J. Symons (Eds.), *Pain in children and adults with developmental disabilities* (pp. 121-135). Baltimore, Maryland: Brookes.

Kennedy, C.J., & Thompson, T. (2000). Health conditions contributing to problem behavior among people with mental retardation and developmental disabilities. In M. Wehmeyer & J. Patten (Eds.), *Mental retardation in the 21st century* (pp. 211-231). Austin, TX: PRO-ED.

Lauger, K., Cornelius, N., & Keedy, W. (2005). Behavioral features of CHARGE syndrome: Parents' perspectives of three children with CHARGE syndrome. *American Journal of Medical Genetics, 133A,* 291-299.

Lotan, M., Moe-Nilssen, R., Ljunggren, A.E., & Strand, L.I. (2010). Measurement properties of the non-communicating adult pain checklist (NCAPC): A pain scale for adults with intellectural and developmental disabilities, scored in a clinical setting. *Research in Developmental Disabilities, 31,* 367-375.

Nicholas, J. (2010). Experiencing pain. In T.S. Hartshorne, M.A. Hefner, S.L.H. Davenport, & J. Thelin (Eds.), *CHARGE syndrome* (pp. 339-351). Plural Publishing.

McGrath, P.J., Rosmus, C., Camfield, C., Campbell, M.A., & Hennigar, A. (1998). Behaviors 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 and Development, 30,* 353-360

Oberlander, T.F., & Symons, F.J. (2006). An introduction to the problem of pain in developmental disability. In T.F. Oberlander & F.J. Symons (Eds.), *Pain in children and adults with developmental disabilities* (pp.1-4). Baltimore, Maryland: Brookes.

Pagon, R.A., Graham, J.M., Zonana, J., & Yong, S.L. (1981). Coloboma, congential heart disease, and choanal atresia with multiple anomalies: CHARGE association. *Journal of Pediatrics, 99,* 223-227.

Palermo, T.M. (2000). Impact of recurrent and chronic pain on child and family daily functioning: A critical review of the literature. *Journal of Developmental and Behavioral Pediatrics*, 21, 58-69.

Phan, A., Edwards, C.L., & Robinson, E.L. (2005). The assessment of pain and discomfort in individuals with mental retardation. *Research in Developmental Disabilities, 26,* 433-439.

Peltokorpi, S. & Huttunen, K. (2008). Communication in the early stage of language development in children with CHARGE syndrome. *The British Journal of Visual Impairment, 26,* 24-49.

Porter, L.S., Davis, D., & Keefe, F.J. (2007). Attachment and pain: Recent findings and future directions. *Pain*, 128, 195-198.

Power, T.J., Heathfield, L.T., McGoey, K.E., & Blum, N.J. (1999). Managing and preventing chronic health problems in children and youth: School psychology's expanded mission. *School Psychology Review*, 28(2), 251-263.

Recchia, S.L. (1997). Social communication and response to ambiguous stimuli in toddlers with visual impairments. *Journal of Applied Developmental Psychology, 18,* 297-316.

Salem-Hartshorne, N., & Jacob, S. (2004). Characteristics and development of children with CHARGE association/syndrome. *Journal of Early Intervention, 26,* 292-301.

Salem-Hartshorne, N. & Jacob, S. (2005). Adaptive behavior in children with CHARGE syndrome. *American Journal of Medical Genetics, 133A,* 262-267.

Sobsey, D. (2006). Pain and disability in an ethical and social context. In T.F. Oberlander & F.J. Symons (Eds.), *Pain in children and adults with developmental disabilities* (pp.19-39). Baltimore, Maryland: Brookes.

Sokol, D., Sokol, S., & Sokol, C. (1985). A review of non-intrusive therapies used todeal with anxiety and pain in the dental office. *Journal of the American Dental Association, 110,* 217-222.

Stallard, P., Williams, L., Lenton, S., & Velleman, R. (2001). Pain in cognitively impaired, non- communicating children. *Archives of Disease in Childhood, 85,* 460-462.

Stratton, K.K. & Hartshorne, T.S. (2010). *Identifying pain in CHARGE syndrome*. (Unpublished master's thesis). Central Michigan University, MI.

Symons, F.J., & Danov, S.E. (2005). A prospective clinical analysis of pain behavior and self- injurious behavior. *Pain, 117,* 473-477.

Symons, F.J., Harper, V.N., McGarth, P.J., Breau, L.M., & Bodfish, J.W. (2009). Evidence of increased non-verbal behavioral signs of pain in adults with neurodevelopmental disorders and chronic self-injury. *Research in Developmental Disabilities, 30,* 521-528.

Taddio, A., & Oberlander, T.F. (2006). Pharmacological management of pain in children and youth with significant neurological impairments. In T.F. Oberlander & F.J. Symons (Eds.), *Pain in children and adults with developmental disabilities* (pp.193-211). Baltimore, Maryland: Brookes.

Tedstone, J.E., & Tarrier, N. (2003). Posttraumatic stress disorder following medical illness and treatment. *Clinical Psychology Review, 23,* 409-448.

Twycross, A., Mayfield, C., & Savory, J. (1999). Pain management for children with special needs: a neglected area? *Paediatric Nursing, 11,* 43-45.

Thelin, J.W., Hartshorne, T.S., & Hartshorne, N.S. (1999). Audiologic and educational issues in CHARGE syndrome. *Journal of Educational Audiology, 7,* 34-41.

Turk, D.C. & Melzack, R. (2001). *Handbook of pain assessment—Second edition.* New York, NY: Guilford Press.

Vaalamo, I., Pulkkinen, L., Kinnunen, T., Kaprio, J., & Rose, R.J. (2002). Interactive effects of internalized and externalizing problem behaviors on recurrent pain in children. *Journal of Pediatric Psychology, 27,* 245-257.

Van der Kolk, B.A. (2005). Developmental trauma disorder: Toward a rational diagnosis for children with complex trauma histories. *Psychiatric Annals, 35,* 401-408.

Van Dijk, J.P.M., & de Kort, A. (2005). Research Letter: Reducing challenging behaviors and fostering efficient learning of children with CHARGE syndrome. *American Journal of Medical Genetics, 133A,* 273-277.

Verloes, A. (2005). Updated diagnostic criteria for CHARGE syndrome: a proposal. *American Journal of Medical Genetics, 133A,* 306-308.

Vissers, L. E., van Ravenswaaij, C. M., Admiraal, R., Hurst, J. A., de Vries, B. B., Janssen, I.M., … van Kessel, A.G. (2004). Mutations in a new member of the chromodomain gene family cause CHARGE syndrome. *Nature Genetics, 36,* 955-957.

Von Baeyer, C.L., & Spagrud, L.J. (2007). Systematic Review of observational (behavioral) measures of pain for children and adolescents aged 3 to 18 years. *Pain, 127,* 140-150.

Waters, E., Davis, E., Mackinnon, A., Boyd, R., Graham, H.K., Kai Lo, S., … Reddihough, D. (2007). Psychometric properties of the quality of life questionnaire for children with CP. *Developmental Medicine & Child Neurology, 49,* 49-55.

Wasserman, J.D. & Bracken, B.A. (2003). Psychometric characteristics of assessment procedures. In J.R. Graham & J.A. Naglieri. (Eds.), *Handbook of psychology: Assessment psychology Volume 10* (43-66). Hokoken, NJ: Wiley.

Waters, E., Maher, E., Salmon, L., Reddihough, D., & Boyd, R. (2005). Development of a condition-specific measure of quality of life for children with cerebral palsy: empirical thematic data reported by parents and children. *Child: Care, Health & Development, 31,* 127-135.

Wulffaert, J., Scholte, E.M., Dijkxhoorn, Y.M., Bergman, J.E.H., van Ravenswaaij-Arts, C.M.A., van Berckelaer-Omnes, I.A. (2009). Parenting stress in CHARGE syndrome and the relationship with child characteristics. *Journal of Development and Physical Disabilities, 21,* 301-313.