Overlapping Pain and Psychiatric Syndromes

GLOBAL PERSPECTIVES

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Effective Management of Pain in Autism Spectrum Disorder and Intellectual Disability

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INTRODUCTION

Research has shown that people with intellectual disability (ID) or autism spectrum disorder (ASD) have markedly higher rates of chronic medical conditions than do people in the general population.[1] Relatively little empirical knowledge is available to guide our understanding and treatment of pain among people with ASD. Compounding this paucity of knowledge are notions that persons with ASD are insensitive or indifferent to pain.

Pain behavior can be ambiguous, leading to confusion and highly subjective assessments that present a tremendous challenge for clinicians, researchers, patients, and their families. Even when pain-specific behaviors are evident, they have been regarded as altered, blunted, or confused with other sources of generalized stress or arousal, or misinterpreted as indicative of general emotional stress or autonomic dysregulation (rage behaviors). This notion has been reinforced by standardized texts[2] reporting that autism is associated with "a high threshold for pain." Our understanding of pain in ASD is very limited and to a great extent based on anecdotal reports and clinical studies derived from heterogeneous populations. Yet, these individuals are often undertreated for pain or not treated for it at all because health care professionals either don't know how to assess their pain or don't recognize that certain nonverbal behaviors can be the expression of pain.

While ASD alters typical forms of communication, everyday interests, and behaviors, there are no data to support the commonly held belief that persons with an ASD experience pain any less frequently or severely than others.

Unfortunately, few studies have addressed the issue of pain assessment and treatment in people with ID or ASD. Using retrospective chart reviews, Bosch et al. considered 25 patients who engaged in self-injurious behavior and found that seven (28%) had previously undiagnosed medical conditions that could cause pain or discomfort. After these conditions were treated, self-injurious behavior decreased in six of the seven (86%).[3] The investigation of several related phenomena may lead to an understanding of possible relationships between neurodevelopment and the pain system. Together, such disparate findings are difficult to reconcile but might suggest that altered pain sensitivity in ASD may be related to differences in mode of pain expression or levels of function, coupled with an altered endogenous biological capacity to mount a nociceptive response.

EXPLORING THE EXPERIENCE OF PAIN IN AUTISM SPECTRUM DISORDER AND INTELLECTUAL DISABILITY

To date, few studies have been conducted on sensitivity to pain stimulation and pain expression in this population.

Hyporeactivity

Individuals with ASD have been described as having reduced sensitivity to pain,[4] indifference to pain,[5] and a high pain threshold.[2] Thus, these reports of altered pain sensation were based on anecdotal observations and clinical impressions.[6]

However, some experimental studies also found this hyporeactivity. Tordjman et al.[7] reported a decrease in behavioral reactivity in children with ASD compared with a control group while performing a venipuncture. Similarly, Pernon and Rattaz[8] found that children with ASD have a decrease in facial reactivity during painful stimulation compared with children without ASD.

Hyperreactivity

Tordjman et al.,[7] in their study, noted reactive behaviors immediately after venipuncture despite a decrease in motor reactivity at the time of the painful procedure. Nader et al.[5] observed greater facial reactivity in 3- to 7-year-old children with ASD during the venipuncture compared with children without ASD.

However, the use of bundling to aid in the safety of the procedure in that study limited the interpretation of this finding.[5]

Sensitivity Threshold

There has been very little research on the topic of sensitivity threshold. Clinicians and families report a high sensitivity for pain. Objective observational measures should be systematically used in studying sensitivity thresholds in ASD patients.[9]

Self-Aggression

In the clinical setting, problem behavior of a self-aggressive nature is not uncommon. Some researchers consider that self-aggression is more related to the stress associated with the examination than to pain. The link between self-harm and pain has not been clearly established. The exacerbation of self-injurious behaviors (such as head banging, hitting or biting oneself, or throwing oneself against hard objects) in people with severe cognitive impairment can be an indication of pain. Thus, the role of discomfort or pain in self-injurious behavior deserves a careful evaluation.[10]

Sleep Disorders

Sleep disorders are frequent comorbidities of ASD, and their prevalence ranges from 50 to 80% of people with autism. Tudor et al. show that the experience of pain in these people is a predictor of sleep disorders, including disruptions, the occurrence of parasomnia, and nocturnal awakenings.[11]

Gastrointestinal Tract

Gastrointestinal problems are significantly overrepresented in ASD and can often be related to problem behaviors, sensory responsivity, sleep disorders, rigid-compulsive behaviors, aggression, anxiety, and irritability.[12] In individuals with ASD, atypical presentation of common gastrointestinal problems can include the emergence or intensification of seemingly autistic behaviors such as self-harm, irritability, aggression, strange posture or movements, and pain.[13]

UNDERSTANDING THE NATURE OF PAIN IN AUTISM SPECTRUM DISORDER AND INTELLECTUAL DISABILITY

Several studies have reported insensitivity to pain in the autistic population compared with control groups, but the contribution of other factors that may partially explain the results is lacking. Using a sociocommunicative perspective, Craig[14] has offered an alternative explanation for the pain insensitivity in people with ASD. The altered perception of others derives from inadequate communication skills and social relatedness.

Direct links between the altered neurological substrate that underlies ASD and nociceptive systems remain elusive. Many hypotheses have been put forward to explain the difference between feeling and expressing pain in people with autism:

- **Neurochemical hypotheses with dysfunction of the opioid or serotonin system, but no conclusive data exists.** Opioid hyperfunction may account for reported pain insensitivity and may be linked to a variety of factors, including a genetic-related opioid system dysfunction that could lead to overproduction, deficient degradation, abnormal feedback, or messenger mechanisms. Research findings addressing opioid system function in ASD have been mixed. The serotoninergic hypothesis has been little studied, although selective serotonin inhibitors are used to reduce certain symptoms such as anxiety and stereotyped behaviors in ASD.
- **Sensory abnormalities in autistic people.** These anomalies concern all sensory modalities and are understood in terms of sensory hyporeactivity or hyperreactivity, decrease in the threshold of stimulus discrimination, or difficulty in modulating sensory information.
- **Abnormal social relationships, adaptive communication, and difficulties in regulating the expression of negative emotions** may explain the atypical pain expression.[15] In addition, Buie et al.[16] provide integrated modeling that takes into account the different hypotheses involving intrapersonal factors (sensory alterations, self-harm, language disorders) and environmental factors (the observer's ability to understand the message) in the perception and expression of pain.

These specific aspects of pain perception and expression should be taken into account when considering the particularities of autistic functioning such as difficulties in modulating sensory influx, somatosensory alterations, communication alterations, and abnormal social relationships. These elements have implications for the management of pain in autistic people, and more broadly for the management of problem behaviors. The most challenging task is to assess and interpret the painful symptomatology and to consider other medical conditions as causes.

PAIN ASSESSMENT

Until clinical research is conducted using a rigorous methodology, the atypical expression of pain raises the question of how to assess and manage pain in people with ASD, particularly when there are major communication disorders. The pain can be underestimated and therefore undertreated. Together, given altered facial emotional expression, social responsiveness, and appropriate use of language, it is likely that typical ASD expressions of pain are altered. In the face of acute pain, a person with ASD might not cry, use appropriate verbal communication, or seek comfort from a caregiver, leading to a perception that the person with ASD is not experiencing pain. It is important to know the patient's social network, such as family members, caregivers, nurses, and others, in order to interpret the manifestation (pleasure/displeasure, comfort/discomfort) of pain. It is recommended to use all possible means of communication (e.g., pictogram, digital tablets, computer, sign language).

Pain assessment is commonly achieved through self-report, observational, and/or proxy methods. Impaired communication skills may make self-report difficult for people with ASD.[14,17,18] Unfortunately, observational pain assessment can also be difficult because of idiosyncratic behaviors associated with ASD and/or cognitive impairment (atypical vocalizations, facial expressions) that may result in inaccurate estimates of pain by those unfamiliar with the individual's typical behavioral responses.[19] Consequently, different observers/caregivers may interpret pain behaviors differently.[20]

It is worth noting that certain behaviors may suggest an organic pathology or pain[21] such as previously acquired skills abilities, a sudden change in behavior, anger and opposition, irritability, sleep disorders, tapping the throat, eating disorders, teeth grinding, making faces, frowning, tics, self-mutilation, constant ingestion, vocalization, and moaning.

Given the complexity of these atypical manifestations of pain, Barthelemy Durand Hospital has set up care protocols tailored to individuals with ASDs. The protocols take into account the environment, sensorial approach, patience, and advance planning of the routine care. Systematic assessment of pain with methods tailored to each case is used for specific and personalized management.[21,22]

The semiological signs of pain expression in ASD identified are aggressiveness, self-harm, screaming without particular cause, sleep disorders, unexplained violent explosion, and repetitive stereotyped movements in the painful area. These signs should be considered to indicate a painful condition and require a systematic investigation into the etiology of pain.[21,23,24]

Pain Comorbidity

Comorbidities are frequent in this population, the most significant are the following:

- **Gastrointestinal symptoms**
  Gastrointestinal symptoms are significantly over-represented in ASD and are often related to behavior problems, sensory over-responsivity, dysregulated sleep, aggression, anxiety, and irritability.[13] Several studies have shown a high prevalence of gastrointestinal symptoms in children with ASD compared with children without ASD, with an odds ratio of 4.5.[25] In recent years, there has been increasing recognition of gastrointestinal comorbidities—both functional bowel problems and pathological findings—among people with ASD, including diarrhea, constipation, gastroesophageal reflux, gastritis, duodenitis, and colitis. In ASD, atypical presentation of common gastrointestinal problems can include the emergence or intensification of self-harm, irritability, aggression, strange posturing or movements, and pain. When abdominal pain or discomfort is the first event and the appropriate medical treatment is effective, the disruptive behavior may diminish.
- **Epilepsy**
  The prevalence of epilepsy is increased in ASD and is even higher in those with co-occurring ID, ranging from 5 to 46% in various studies.[26] The clinician should be careful when individuals with ASD present a family history of seizures, multiple febrile seizures, a first-time afebrile seizure, sleep disturbances, ID with or without focal neurological abnormalities, or a history of regression. Pain has been described related to epileptic seizures, most often partial and atypical.
- **Dental disease**
  Dental conditions should be considered as a source of discomfort or pain, particularly in children presenting with irritability or agitation. Cavities, dental infection, and dental erosion are common sources of pain.

- **Menstrual pain or premenstrual syndrome**
  Some medical problems that emerge in adolescence may be difficult to identify in youths with ASD, particularly those who have communication difficulties. Migraine headaches may emerge during early to mid-adolescence. Careful attention should be paid to clinical manifestations of headache, including squeezing of the head or increased sensitivity to sound and light.
  Menstrual pain is another common cause of distress in adolescent girls with ASD and is often undertreated. Premenstrual dysphoric disorders should be considered when mood fluctuation occurs just before menstruation. Clinicians should be attentive to the possibility of epilepsy during adolescence and even extending into adulthood.

Other Sources of Pain

Many other conditions can cause pain or irritability, such as pressure sores, otitis, sinusitis, or fractures and dislocations. During the clinical examination, it is imperative to detect and manage them to avoid disruptive behaviors.

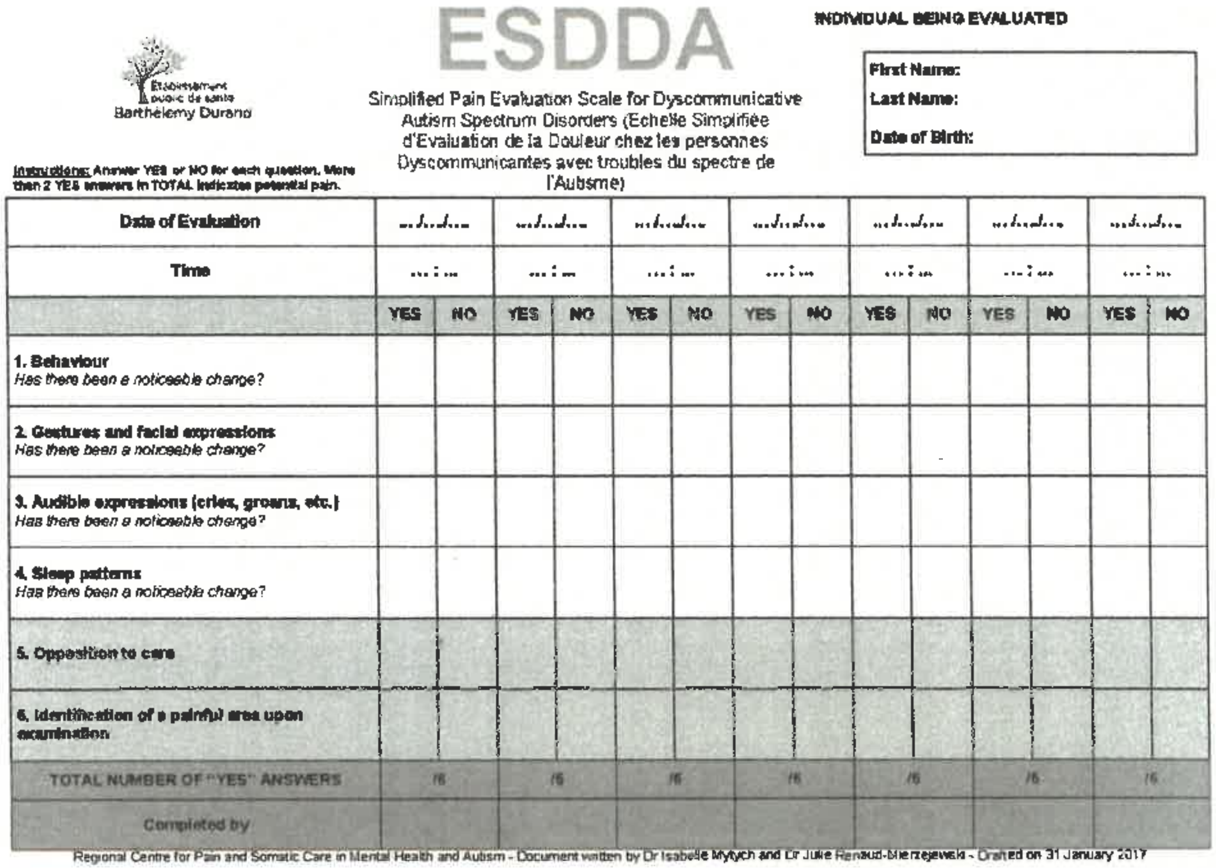
PAIN ASSESSMENT TOOLS

The identification and assessment of pain is the responsibility of everyone, including those close to the person with ASD or ID, including family members, caregivers, and professionals. If pain is detected, the assessment will be challenging and require knowledge and careful observation.

The assessment must be carried out using appropriate tools. It is important to keep in mind how changes in nonverbal and social behavior and inability to develop social relationships affect the standard use of these pain assessment tools. Poor eye contact or lack of social expression could negatively influence measurements. Tools developed to identify pain in people with intellectual disabilities[27,28] may provide ways to assess pain in people with severe cognitive impairment. Thus, we can consult the GED DI (pain/intellectual disability assessment grid), the EDAAP (assessment of pain in adolescents and adults with multiple disabilities), or the ESD (a pain assessment tool for individuals with communication disorders, the EDAAP adapted for ASD).

Barthelemy Durand Hospital developed a tool to help identify potentially acute pain. This tool: ESDDA: simplified pain assessment tool for people with communication difficulties in ASD,[23] is provided in Table 26.1

Table 26.1 USER GUIDE TO THE ESDDAA PAIN SCALE



Barthélemy Durand

ESDDA

Simplified Pain Evaluation Scale for Dyscommunicative Autism Spectrum Disorders (Echelle Simplifiée d'Evaluation de la Douleur chez les personnes Dyscommunicantes avec troubles du spectre de l'Autisme)

INDIVIDUAL BEING EVALUATED

First Name:
Last Name:
Date of Birth:

Instructions: Answer YES or NO for each question. More than 2 YES answers in TOTAL indicates potential pain.

Date of Evaluation	…/…/…		…/…/…		…/…/…		…/…/…		…/…/…		…/…/…		…/…/…	
Time	…:…		…:…		…:…		…:…		…:…		…:…		…:…	
	YES	**NO**	**YES**	**NO**	**YES**	**NO**	**YES**	**NO**	**YES**	**NO**	**YES**	**NO**	**YES**	**NO**
1. Behaviour *Has there been a noticeable change?*														
2. Gestures and facial expressions *Has there been a noticeable change?*														
3. Audible expressions (cries, groans, etc.) *Has there been a noticeable change?*														
4. Sleep patterns *Has there been a noticeable change?*														
5. Opposition to care														
6. Identification of a painful area upon examination														
TOTAL NUMBER OF "YES" ANSWERS	/6		/6		/6		/6		/6		/6		/6	
Completed by														

Regional Centre for Pain and Somatic Care in Mental Health and Autism - Document written by Dr Isabelle Mytych and Dr Julie Renaud-Mierzejewski - Drafted on 31 January 2017

This scale defines six items: behavior, gestures and facial expression, audible expressions (cries, groans), sleep patterns, opposition to care, and identification of a painful area on examination. More than two positive answers in total indicate a potential pain. This tool can be used by health care as well as non-health care staff, including family members, caregivers, educators, social workers, and others.

Approaches focused on sensitive and specific methods for measuring nonverbal manifestations of pain and behavioral reactivity to procedural pain have been investigated, but the clinical usefulness of these methods is yet to be proved.[29]

PAIN MANAGEMENT IN AUTISM SPECTRUM DISORDER AND INTELLECTUAL DISABILITY

The management of pain must include the identification, when possible, of the painful pathology, its evaluation, and the preparation of treatment plans aimed at reducing problem behaviors. But, even with a careful history and specific approaches investigating irritability, identifying specific sources of pain may remain difficult. A pain-related diagnosis may not always be possible; however, even after a careful empiric evaluation, an empiric medication trial and careful ongoing assessment may be the only available management options.

Therapeutic options may be based on pharmacological and non-pharmacological options.[28]

Pharmacological Treatment

Analgesic treatments are prescribed according to the modalities of use in general population. However, special precautionary measures must be taken into account:

- Oral or transdermal routes of drug administration are preferred. Intramuscular injections should be avoided, especially in the event of prolonged treatment (muscle mass may be reduced, atypical stress).
- Because of bowel movement problems and the frequency of constipation, prolonged use of opioids should be avoided and take preventive measures.
- Topical anesthetic creams may be recommended for care-induced pain (e.g., for blood sampling), or an equimolar mixture of nitrogen protoxide and oxygen may be used for clinical pain assessment examinations.

Problem behavior in ASD may be the primary or sole symptom of an underlying illness causing pain, which can be acute or chronic. When pain or discomfort is a main complaint, the implementation of inappropriate treatment, such as psychotropic medication, or of aberrant behavioral interventions, including hypostimulation or isolation rooms, is ineffective and may worsen the clinical condition.

Nonpharmacological Options

Very few studies have been conducted on the effectiveness of nonpharmacological approaches to pain management in ASD patients. Physical approaches such as massage, touch, and hot and cold physiotherapy techniques can be very helpful. Other therapeutic strategies, such as music, certain osteopathic techniques, and adaptive physical activity, can be offered.

Some video entertainment tools can also be useful (digital tablets, movies, video games).

CONCLUSION

Medical comorbidities and pain can be difficult to recognize in patients suffering from ASD or ID. The failure to identify medical illness and pain is due in part to communication impairments and sometimes aberrant symptomatology. Widespread underdiagnoses and access barriers to appropriate health care for ASD are the result of commonly held beliefs that aberrant behaviors and symptoms are "just a part of autism."

Understanding the atypicality of autistic functioning in relation to the specific characteristics of pain perception and expression in people with ASD or ID must break with the long-standing assumption that these people are insensitive to pain. Behavioral changes should be a sign that there is an underlying experience of pain. The assessment of the pain and its treatment should be a priority. This assessment must be personalized, taking into account the problem behavior and its frequency, intensity, and duration, as well as its context and associated events. Pain in people with ASD, who are vulnerable, must be taken into account and treated as in any other individual. Leaving pain untreated clearly results in health inequalities and constitutes a gross injustice to people with ASD.

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