Critical Review

Behavioral Pain Indicators in People With Intellectual Disabilities: A Systematic Review

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Abstract: People with intellectual disabilities (IDs) have a higher risk of painful medical conditions. Partly because of the impaired ability to communicate about it, pain is often undertreated. To strengthen pain assessment in this population, we conducted a systematic review to identify behavioral pain indicators in people with IDs by using Embase, PubMed, PsycINFO, CINAHL, and Cochrane. Inclusion criteria were 1) scientific papers; 2) published in the last 20 years, that is, 1992 to 2012; 3) written in English, 4) using human subjects, 5) intellectual disabilities, 6) pain, 7) behavior, and 8) an association between observable behavior and pain experience. From 527 publications, 27 studies were included. Pain was acute in 14 studies, chronic in 2 studies, both acute and chronic in 2 studies, and unspecified in 9 studies. Methodological quality was assessed with the Mixed Methods Appraisal Tool. Of the 14 categories with behavioral pain indicators, motor activity, facial activity, social-emotional indicators, and nonverbal vocal expression were the most frequently reported. Most of the behavioral pain indicators are reported in more than 1 study and form a possible clinical relevant set of indicators for pain in people with IDs. Determination of a behavioral pattern specific for pain, however, remains a challenge for future research.

Perspective: This review focuses on categories of behavior indicators related to pain in people with IDs. The quality of evidence is critically discussed per category. This set of indicators could potentially help clinicians to recognize pain in this population, especially when unique individual pain responses are also identified.

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Key words: Intellectual disabilities, pain assessment, behavior, detection.

The prevalence of pain is likely to be high in people with intellectual disabilities (IDs) because comorbidity is common and medical procedures such as surgery are often needed. Pain or discomfort is caused by medical conditions such as musculoskeletal problems, gastroesophageal reflux disease, and oral diseases and may impact the daily lives of people with IDs. Caregivers’ reports suggest that 14 to 15% of adults with IDs have chronic pain, the average duration being 6 years. As certain painful conditions such as arthritis occur more often in older adults with IDs than in younger adults, and as improved health care increases the life expectancy of the IDs population, the clinical relevance of identifying pain in adults with IDs has never been higher. The frequent experience of severe pain in children with IDs and the interference of pain with their daily life illustrate the urgent need to detect pain also in children with IDs. Thus, pain detection

Received December 13, 2012; Revised April 12, 2013; Accepted April 25, 2013.
The PhD project of N. C. de Knegt (“Pain and Cognition in Adults With Down Syndrome”) is possible due to funding of the following Dutch organizations: Fonds NutsOhra, Fonds Verstandelijk Gehandicapten, Innovatiekring Zorgverzekeraars, and Alzheimer Nederland. There are no conflicts of interest to report for any of the authors. Supplementary data accompanying this article are available online at www.jpain.org and www.sciencedirect.com.
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1526-5900/$36.00
© 2013 by the American Pain Society
http://dx.doi.org/10.1016/j.jpain.2013.04.016
is relevant for the entire IDs population, regardless of age.

Despite this relevance, pain seems to be undertreated in people with IDs. Because of the cognitive difficulties with reflecting on their own pain experience, self-report of pain is often difficult. The fact that many people with IDs are nonverbal complicates any communication about the pain. Self-report of pain may also be avoided by people with IDs for fear of disease or death. Such communication difficulties hamper an adequate prescription of analgesics and palliative care to people with IDs. Therefore, to prevent undertreatment of pain in patients with IDs, it is important to find an alternative method for self-reported pain assessment. Because behavioral observation is recommended for pain assessment in people with communication difficulties, such as dementia (International Association for the Study of Pain17; American Society for Pain Management Nursing18), we argue that it will also strengthen pain assessment in people with IDs.

The literature suggests that behavioral observation of pain is not a method without difficulties, because behavioral pain indicators in people with IDs are not well recognized by health care professionals. There are various explanations for this problem. First, people with IDs may display typical behavioral pain indicators such as facial activity and moaning even in the absence of pain. Second, it is not yet clear whether self-injurious behavior in people with IDs is a behavioral pain indicator. Third, the various IDs syndromes could have a specific profile of increased or decreased pain experience resulting from unique neuropathology in white and gray matter. Fourth, multiple comorbidity in people with IDs—for example, a combination of epilepsy and a psychiatric disorder—complicates the identification of pain-related symptoms. And fifth, behavioral pain indicators could differ between acute and chronic pain. Acute pain is associated with indicators such as tears, not moving, and squeezing the eyes shut, whereas chronic pain in people with IDs may be associated with indicators such as moaning, eating less, and not cooperating.

Because of these difficulties and the importance to detect pain in the IDs population, more information is needed about behavioral pain indicators in both children and adults with IDs. The goal of the present systematic review was to define behavioral pain indicators in people with IDs and to evaluate the methodological quality of studies that addressed this topic. Behavioral pain indicators imply a set of observable verbal or nonverbal behaviors that could be considered an expression of, and a reaction to, physical pain subjectively experienced by the individual. We discriminated between behavioral indicators for acute pain and chronic pain. Acute pain persists within the phase of 3 months following an injury and needs immediate treatment to prevent the development into chronic pain. Chronic pain persists past the healing phase of 3 months following an injury, resulting in reduced treatment possibilities and a psychosocial impact on daily living.

Methods

Searching

A systematic literature search was conducted on October 9, 2012, using inclusion and exclusion criteria that are described in Table 1. Five databases were used: Embase (348 results after first search), PubMed (131 results after first search), PsycINFO (66 results after first search), CINAHL (21 results after first search), and Cochrane (5 results after first search). The exact MeSH terms differed slightly between the search strategies that were used in these databases, and it was considered neither informative nor clarifying to write out these 5 search strategies. As a relevant example for this Methods section, the search strategy was illustrated for the database with which most of the papers were retrieved after the first search. Thus, although all the 5 databases and relating search strategies were included in this systematic search, only the MeSH terms were specified for Embase (348 results). (MeSH terms are called “Emtree terms” in Embase.) We combined the following components (1–3) and limited the results to humans, Embase, and English. “Exp” means exploded, that is, extended; Emtree term “ab” means abstract, and “ti” means title.

1. Intellectual Disability

(“mental deficiency”/exp OR “mental deficiency”:ab,ti OR “mental retardation”:ab,ti OR “intellectual disabilities”:ab,ti OR “intellectual disability”:ab,ti OR “Down syndrome”:ab,ti)

2. Pain

“pain”:exp OR “pain assessment”:exp OR “pain perception”:exp OR “analgiesia”:exp OR pain*:ab,ti OR discomfort*:ab,ti.

3. Behavior

“behavior”:exp OR “observation”:exp OR “cues”:exp OR “behavior”:ab,ti OR “observation”:ab,ti OR “cues”:ab,ti.

Selection

Definition

According to the fourth text-revised version of the Diagnostic and Statistical Manual of Mental Disorders,1 borderline IDs refers to an IQ level of 70 to 85, mild IDs refers to an IQ level of 50 to 70, moderate IDs refers to an IQ level of 35 to 50, severe IDs refers to an IQ level of 20 to 35, and profound IDs refers to an IQ level below 20.

Trial Flow

In Fig 1, we present a flow diagram, according to the PRISMA guidelines,30 that summarizes the results of our literature search. After the removal of duplicates, 527 publications were initially identified. The entire process of selection of titles, abstracts, and full texts was executed by the first author (N.C.d.K.) strictly on the basis of the inclusion and exclusion criteria described in Table 1. During the writing of this review, 1 study was added on the basis of a personal communication from an expert in the field. The selection process...
resulted ultimately in 27 studies included for qualitative synthesis.

**Quality Assessment**

The criteria of the Mixed Methods Appraisal Tool (MMAT)\(^3\)\(^4\) were used to assess the methodological quality of the 27 eligible studies. The MMAT, described in Supplementary Table 1, was applied by executing the following steps for each study: 1) screening whether there are clear research questions and whether the collected data allow addressing the research question; 2) determining whether the study includes qualitative, quantitative randomized controlled, quantitative non-randomized, quantitative descriptive, or mixed methods; 3) determining the specific study design within the chosen methodology category; and 4) determining how many of the 4 criteria within the chosen methodology category are met: the score for a study ranges from 25% for 1 criterion met to 100% for 4 criteria met. The 4 criteria were relevant for the corresponding methodology category: for example, consideration of how findings of qualitative studies relate to the context versus consideration of whether the participants of the groups in quantitative studies are comparable. Because of these specific criteria, a study’s calculated MMAT score, that is, the number of criteria that were met, reflected the study’s methodological quality instead of the methodological category itself.

**Results**

**Study Characteristics**

The characteristics and quality score of the 27 studies are presented in Supplementary Table 2. The 27 included publications consisted of 11 cross-sectional studies without comparison group, 8 cross-sectional analytic studies, 3 case series, 2 qualitative descriptions, 2 case-control studies, and 1 case study. In total, 2,075 people with IDs were included in 26 studies. The number of people with IDs is unclear in 1 study.\(^5\)\(^2\) Most studies (12/27) included both children and adults with IDs, 8 studies included only children, and 7 studies included only adults. For the sake of clarity, we refer to participants below 18 years of age as “children.” Most of the studies included people with unknown levels of IDs (6/27) or severe IDs (5/27). Most of the studies included various levels of IDs: 2 studies involved people with borderline IDs; 7 studies, people with mild IDs; 11 studies, people with severe IDs; and 11 studies, people with profound IDs.

**Methodological Quality**

Most studies (10/27) had an MMAT quality score of 50%, followed by 75% (7/27), 100% (6/27), and 25% (4/27). Details of the methodological aspects of the studies are presented in Supplementary Table 2.

**Study Outcomes**

Table 2 shows the 14 categories of behavioral pain indicators, ordered by the total number of studies reporting them and specified according to source of information and nature of pain. Each study could cover more than 1 behavioral pain indicator category. The sources of information were expert opinion, that is, opinion of health care professional or parent, and observation by caregiver, that is, health care professional or parent, or researcher. The nature of pain was acute pain, chronic pain, and both acute and chronic pain; some studies did not specify the nature of the pain. Pain was acute in 14 studies, chronic in 2 studies, both acute and chronic in 2 studies, and unspecified in 9 studies. The categories of behavioral pain indicators

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**Table 1. Inclusion Criteria and Exclusion Criteria for Systematic Search**

<table>
<thead>
<tr>
<th>Inclusion Criteria</th>
<th>Exclusion Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Articles (all types of study design described in the articles were included [eg, cross-sectional, case-control, case series])</td>
<td>Other publication types than articles, eg, reviews or published comments/replies on articles</td>
</tr>
<tr>
<td>Written in English language</td>
<td>Written in a language other than English</td>
</tr>
<tr>
<td>Human studies</td>
<td>Animal studies</td>
</tr>
<tr>
<td>Intellectual disability (synonyms in Methods section of papers that were included: mental retardation, intellectual and developmental disabilities)</td>
<td>Other conditions (eg, autism, cerebral palsy, or epilepsy) without IDs</td>
</tr>
<tr>
<td>Pain (eg, period pain, abscessed teeth, duodenal ulcer)</td>
<td>Mere distress or discomfort, especially without situation that most likely is causing pain (eg, common dental procedure compared to the likely painful postoperative period or vaccination)</td>
</tr>
<tr>
<td>Behavior: observed, based on expert opinion, or self-reported Association between observable behavior and pain experience (eg, observation list with behavioral pain indicators used during vaccination)</td>
<td>Congenital insensitivity to pain/hereditary sensory neuropathy/heriteditary sensory and autonomic neuropathy</td>
</tr>
<tr>
<td>No behavior, eg, only measured sensory or pain thresholds</td>
<td>Self-reported pain experience</td>
</tr>
<tr>
<td>Sensory pain processing</td>
<td>Self-injurious behavior without painful medical condition (eg, without being certain that period pain is actual present in participants)</td>
</tr>
<tr>
<td>Self-injurious behavior existing prior to the painful medical condition (eg, painful medical condition due to self-injurious behavior)</td>
<td></td>
</tr>
</tbody>
</table>

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will be described next, ordered by the total number of studies reporting them and thus corresponding to the order in Table 2.

**Categories of Behavioral Pain Indicators**

**Motor Activity**

Two studies, acute pain, 2 studies both acute and chronic pain, and 7 studies an unspecified nature of pain. The results were derived from studies at all levels of methodological quality: 5 studies had a 100% quality score, 3 studies had a 75% quality score, 5 studies had a 50% quality score, and 2 studies had a 25% quality score.

The “motor activity” category reflects increase in physical activity, passivity, and/or posture of the body. Parents and nursing staff rated various physical (in)activities as important for pain recognition in people with IDs. Examples are not moving; not using a body part; flinching or moving a body part away; being sensitive to touch; being stiff, spastic, tense, or rigid; having increased tonic problems; moving the body in a specific way; and protecting, defending, or guarding the hurting body part. For example, resistance to hip examination with chronic hip pain has been observed in a child with IDs. Whether someone who is in pain will react with decreased or increased movement may depend on his or her verbal abilities: absence of moving may be typical for nonverbal children with IDs, whereas kicking has been observed in verbal children with IDs. Verbal children with IDs show more behavioral pain indicators in general than nonverbal children with IDs, such as facial, vocal, and verbal expressions of pain.

Indications for acute period pain or discomfort include lying down in combination with verbal expressions of pain, and holding the abdomen. During admission to the intensive care unit, children with IDs may show more physical movements and less muscle tone than children without IDs, although the lower muscle tone could have been influenced by the fact that the participants with IDs had Down syndrome. After surgery, children with IDs show more behaviors related to body and limbs—for example, gesturing or guarding—and...
<table>
<thead>
<tr>
<th>Behavioral Pain Indicators</th>
<th>Total Number of Studies</th>
<th>Opinion Caregiver or Expert (Reference)</th>
<th>Observation Caregiver or Researcher (Reference)</th>
<th>Acute Pain (Reference)</th>
<th>Chronic Pain (Reference)</th>
<th>Acute and Chronic Pain (Reference)</th>
<th>Not Specified Nature of Pain (Reference)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Motor activity: tonus problems, involuntary movements, protection, posture, defensive reaction, resistance to movement, guarding, lying down, “body and limb,” touching hurting body part, increased or decreased activity level</td>
<td>16</td>
<td>5, 10, 11, 18, 27, 40, 51, 52</td>
<td>7, 8, 12, 15, 24, 33, 41, 48</td>
<td>8, 11, 15, 24, 33, 48</td>
<td>41, 51</td>
<td>7, 10</td>
<td>5, 12, 18, 27, 40, 52</td>
</tr>
<tr>
<td>Facial activity: eg, eyes and mouth. One study did not find a facial response during mild acute pain.</td>
<td>16</td>
<td>5, 10, 27, 40, 51, 52</td>
<td>8, 12, 15, 23, 31, 33, 41, 43, 48, 49</td>
<td>8, 15, 23, 31, 33, 43, 48, 49</td>
<td>41, 51</td>
<td>10</td>
<td>5, 12, 27, 40, 52</td>
</tr>
<tr>
<td>Social-emotional indicators: eg, wanting to be left alone, interest in surroundings, capacity to interact, pointing to or showing an injury, using sign language (“hurt” or “sick”), refusing to do usual activities, more needy of parents, difficult to comfort, signs of fear, distress or anxiety, signs of frustration or irritability, low mood or depression, being upset or stressed</td>
<td>14</td>
<td>5, 10, 11, 13, 18, 22, 27, 40, 51, 52</td>
<td>7, 8, 12, 15</td>
<td>8, 11, 15, 22</td>
<td>51</td>
<td>7, 10</td>
<td>5, 12, 13, 18, 27, 40, 52</td>
</tr>
<tr>
<td>Nonverbal vocal expression: crying, screaming, moaning, groaning, sounds of distress</td>
<td>13</td>
<td>5, 11, 13, 27, 40, 51, 52</td>
<td>8, 12, 15, 36, 48, 49</td>
<td>8, 11, 15, 36, 48, 49</td>
<td>51</td>
<td>-</td>
<td>5, 12, 13, 27, 40, 52</td>
</tr>
<tr>
<td>Physiological indicators: eg, respiration, tears, facial pallor/redness. No evidence for heart rate.</td>
<td>9</td>
<td>5, 22, 27, 40, 52</td>
<td>8, 15, 24, 31</td>
<td>8, 15, 22, 24, 31</td>
<td>-</td>
<td>-</td>
<td>5, 27, 40, 52</td>
</tr>
<tr>
<td>Self-injurious behavior</td>
<td>6</td>
<td>11, 38, 40, 51</td>
<td>4, 36</td>
<td>11, 36</td>
<td>51</td>
<td>-</td>
<td>4, 38, 40</td>
</tr>
<tr>
<td>Verbal expression: location-specific (eg, “My tummy hurts“) or non–location-specific (eg, “I don’t feel well”/“Owie“)</td>
<td>6</td>
<td>11, 13, 22, 51</td>
<td>7, 15</td>
<td>11, 15, 22</td>
<td>51</td>
<td>7</td>
<td>13</td>
</tr>
<tr>
<td>Aggression: throwing objects, striking, destroying furniture, throwing tantrums, acting out, showing signs of anger, challenging behavior</td>
<td>5</td>
<td>11, 13, 51</td>
<td>26, 36</td>
<td>11, 26, 36</td>
<td>51</td>
<td>-</td>
<td>13</td>
</tr>
<tr>
<td>Eating/sleeping: more or less appetite or sleeping than usual</td>
<td>4</td>
<td>5, 22, 27</td>
<td>26</td>
<td>22, 26</td>
<td>-</td>
<td>-</td>
<td>5, 27</td>
</tr>
<tr>
<td>Agitation: restlessness or excessive talking</td>
<td>4</td>
<td>22</td>
<td>15, 24, 36</td>
<td>15, 22, 24, 36</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Daily living skills: eg, self-care</td>
<td>2</td>
<td>51</td>
<td>7</td>
<td>-</td>
<td>51</td>
<td>7</td>
<td>-</td>
</tr>
<tr>
<td>“Exaggeration of usual symptoms of the disability”</td>
<td>1</td>
<td>22</td>
<td>-</td>
<td>22</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Response to analgesics</td>
<td>1</td>
<td>22</td>
<td>-</td>
<td>22</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Stereotyped movements: eg, hands flapping</td>
<td>1</td>
<td>-</td>
<td>15</td>
<td>15</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Total (1 study can cover several indicators)</td>
<td>98</td>
<td>54</td>
<td>44</td>
<td>46</td>
<td>10</td>
<td>7</td>
<td>35</td>
</tr>
</tbody>
</table>

NOTE: Studies are specified per source of information (opinion or observation) and per nature of pain (acute, chronic, acute and chronic, or not specified). Reference 25 is missing because of unspecified study outcomes. Caregiver = parent or health care staff member such as nurse. Opinion = eg rating the importance of items or developing a checklist based on experience. Observation = Facial Action Coding System, observation lists, or case series. Acute pain = pain lasting ≤3 months. Chronic pain = pain lasting >3 months. Not specified nature or pain = duration of pain is unclear or not mentioned or examined.
changed activity level than before the surgery. A changed activity level could be either less activity or more activity. Physical movements, activity level, and muscle tone are rated by primary caregivers as behavioral pain indicators for people with IDs. In particular, a decreased activity level was reported, such as being slower in doing usual activities. Caregivers also observed that children with IDs display less motor skills during days with pain than during pain-free days.

**Facial Activity**

These results were derived from studies at all levels of methodological quality: 4 studies had a 100% quality score, 4 studies had a 75% quality score, 6 studies had a 50% quality score, and 2 studies had a 25% quality score.

The “facial activity” category refers to changes in the face, such as eyes closed tight, a tense face, or a deeper nasolabial furrow. These indicators seem to be reliable in people with IDs as indicated by a high interrater agreement and a strong relationship with visual analog scale ratings. A facial expression such as grimacing is observed in both children and adults with IDs who suffer from chronic pain. A specific pattern of an increased intensity of chin raising combined with an increased activity level was found during mild acute pain in adults with IDs.

During acute painful or discomforting situations, children and adults with IDs show more facial activity than during painless situations. Examples of such acute situations are influenza vaccinations, dental cleaning procedures, postoperative pain, and the movement of painful joints for changing clothes. One study shows a contrasting finding; that is, children with Down syndrome display less facial tension after surgery than children without IDs, but the authors state that this difference was not clinically relevant because of the small standardized mean difference. Another study reports no changed facial activity in adolescents with cerebral palsy during an influenza vaccination as compared to a sham condition. The authors suggest that the presence of chronic pain may have influenced the acute pain response. It is important to note that the study contained only 8 participants and that the level of IDs is unclear. A comparison with the other study that used influenza vaccinations is therefore difficult, also because chronic pain was not mentioned in the latter study. As a behavior indicator for acute pain in people with IDs, facial activity seems to be influenced by 2 factors: age and self-injurious behavior. Adults are more likely to show a tense face and a deeper nasolabial furrow than are children. During acute discomforting sensory stimuli, such as pin prick and deep pressure, people who display self-injurious behavior show more facial activity than those without self-injurious behavior, which might suggest a different sensory processing and experience.

There is evidence that the absence of facial expression in people with severe IDs and cerebral palsy may reflect that the person does not experience pain.

**Social-Emotional Indicators**

One study concerned chronic pain, 3 studies acute pain, 2 studies both acute and chronic pain, and 7 studies an unspecified nature of pain. The results were derived from studies at all levels of methodological quality: 4 studies had a 100% quality score, 1 study had a 75% quality score, and 3 studies had a 50% quality score, and 2 studies had a 25% quality score.

The socialization skills of children with IDs, that is, the ability to interact with others and participate in joint activities, could be negatively influenced by pain. Wanting to be left alone and less interaction with others are indications for pain in people with IDs—for example, during acute period pain. In contrast, seeking comfort has also been rated as important to detect pain, although it seems difficult to actually comfort the individual with IDs who is suffering from pain. Moreover, although the category contains contrasting social indicators such as wanting to be left alone versus seeking comfort, caregivers note a change in this category when children with IDs are in pain. Social indicators could be partly related to motor activity—for example, using sign language for “hurt” or “sick.”

Furthermore, a variety of emotional indicators can be used to recognize pain in both adults and children with IDs: irritability, frustration, low mood, depression, being upset or stressed, signs of fear or anxiety, and not being cooperative.

**Nonverbal Vocal Expressions**

The results were derived from studies at all levels of methodological quality: 3 studies had a 100% quality score, 3 studies had a 75% quality score, 6 studies had a 50% quality score, and 1 study had a 25% quality score.

The majority of caregivers value vocalization such as moaning (60–80%) or groaning, crying (60–90%), and screaming as behavioral pain indicators for people with IDs—for example, during acute period pain or discomfort. During acute pain, moaning and groaning were more often observed in adults than in children with a combination of profound IDs and severe or profound motor disabilities. Crying is a behavioral indicator for chronic pain in adults with severe or profound motor disabilities.
Physiological Indicators

Five studies8,15,22,24,31 concerned acute pain and 4 studies5,27,40,52 unspecified nature of pain. The results were derived from studies at all levels of methodological quality: 2 studies had a 100% quality score,15,27 4 studies had a 75% quality score,8,24,31,52 2 studies had a 50% quality score,5,22 and 1 study had a 25% quality score.40

The “physiological indicators” category consists of autonomic bodily symptoms, with individual differences such as facial pallor or redness. Caregivers report that physiological signs are typical pain behaviors for children with IDs.5,40 Children with IDs show more of such signs after surgery than before surgery.8

The relevance of physiological indicators to detect pain in people with IDs, however, is not a consistent finding. Because breathing and gasping were not significantly related with an acute painful stimulus in adults with IDs but some adults did display these behaviors, the broader category of “respiratory irregularities” was formed for an adult pain behavior checklist.24 Parents report that facial pallor is an indication for acute period pain in women with Down syndrome.22 In addition, turning red in the face is a very important symptom in people with profound to severe IDs.52 Tears are observed after surgery in children with IDs, but only in those with verbal abilities.15 Verbal children with IDs show in general more behavioral pain indicators than nonverbal children with IDs, such as facial, vocal, and verbal expressions of pain.15 Heart rate did not change in nonverbal adolescents with cerebral palsy during an influenza vaccination as compared to a sham procedure.31 The usefulness of physiological symptoms as pain indicators varies from 5 to 54%.27,52

Self-Injurious Behavior

The results were derived from studies at all levels of methodological quality: 1 study had a 75% quality score,11 3 studies had a 50% quality score,9,36,51 and 2 studies had a 25% quality score.38,41

According to caregivers, self-injurious behavior could be a typical behavior that people with IDs express when they are in acute40 or chronic51 pain. By means of retrospective interviews, parents indicated physical discomfort as 1 of the 4 possible factors causing self-injurious behavior in their children with IDs.38 Others confirm that self-injurious behavior in adults with IDs could be a reaction to acute pain provoked by untreated medical conditions such as duodenal ulcers.4

Self-injurious behavior is a common cyclical behavior in IDs.35,36 Women with IDs who display self-injurious behavior during menses may benefit from NSAIDs,36 although it remains unclear which of the cyclical behaviors responds to NSAIDs.36 When women with IDs experience period pain or discomfort and they have to do aversive tasks such as making the bed, then the women could display self-injurious behavior to escape from the tasks.11 As these women already used analgesics such as NSAIDs, behavioral and nondrug interventions were needed to reduce self-injurious behavior. In short, the evidence for self-injurious behavior as an indicator for already existing pain in IDs is not yet clear when period pain is concerned.

Verbal Expression

The results were derived from studies at almost all levels of methodological quality: 3 studies had a 100% quality score,7,13,15 1 study had a 75% quality score,11 and 2 studies had a 50% quality score.22,51

Some people with IDs are able to tell that they are experiencing pain,11,13,22,51 both chronic51 and acute pain/discomfort.11,22 A distinction can be made between location-specific words, such as “my stomach hurts,” and non–location-specific words, such as “I don’t feel well.”11,13,15,22 The content and frequency of the verbal reports might be influenced by the interpretation of pain experience by the person with IDs. However, caregivers also observe a relationship between pain and verbal expressions.7,15 Children with IDs show less communication skills, that is, worse language use and understanding, during pain than during a pain-free day.2 The language that they do use during pain—for example after surgery—is recognized by caregivers as a pain behavioral indicator.15 Although some intellectually disabled people with limited communication skills are able to state that they are in pain,22 nonverbal children with IDs are not.15

Aggression

The results were derived from studies at almost all levels of methodological quality: 1 study had a 75% quality score,11 2 studies had a 50% quality score,36,51 and 1 study had a 25% quality score.38

The “aggression” category refers to verbal and nonverbal aggressive behavior. Throwing food, destroying furniture, and striking others could be a reaction to acute pain provoked by dental conditions such as abscessed teeth.26 Pain relief measures, for example,
extraction of the abscessed teeth, resulted in immediate behavioral improvement in 2 persons. More detailed information about the behavior improvement itself was not provided.

Challenging behavior was found to be more prevalent in intellectually disabled adults with chronic pain than those without chronic pain. As the prevalence of verbal abuse and aggression did not differ between those 2 subgroups, the lacking definition of challenging behavior prevents a profound understanding of the evidence for this behavioral pain indicator.

During acute period pain, women with IDs may show aggression or tantrums when they have to execute aversive tasks. However, it is known that cyclical behavior in women with IDs differs from that of women without IDs; tantrums are shown by those with IDs35 and are therefore not necessarily related to pain. Aggression could be related to period pain because cyclical behavioral changes such as aggression improved in many women with IDs who received NSAIDs. It remains unclear, however, which behavior improves as a result of the NSAIDs and women who use analgesics during menses could still display aggression. Therefore, behavioral and nondrug interventions may be needed in addition to analgesics to reduce aggression in women with IDs during menses.

Eating/Sleeping

The results were derived from studies at almost all levels of methodological quality: 1 study had a 100% quality score, 2 studies had a 50% quality score, and 1 study had a 25% quality score. Caregivers mention eating and sleep disturbances as behavioral pain indicators in children and adults with IDs—for example, for acute period pain. In a person with profound IDs and undeveloped speech, poor sleep alarmed the nursing staff that something was wrong, that is, acute painful abscessed teeth. Pain relief by extraction of the abscessed teeth resulted in immediate behavioral improvement. More detailed information about the behavior improvement itself was not provided.

Agitation

Considering that the quality score of the 3 studies was not higher than 75%—that is, 1 study with 75% and 2 studies with 50%—these findings should be considered with caution.

Within the “agitation” category, restlessness and excessive talking were used by caregivers as indicators for acute period pain in women with Down syndrome. In the IDs population, agitation is a behavioral pain indicator of specific importance for adults because many adults were agitated during the acute painful stimulus of an injection. Despite this, women of all ages with IDs could display agitation and restlessness during menses, which may improve by using NSAIDs and thus may be related to period pain.

Daily Living Skills

The quality score of the study about adults was 50% and that of both studies concerning children was 100%.

Adults with IDs who experience chronic pain are limited in their daily living skills, such as exercising and maintaining an independent lifestyle. Children with IDs use fewer daily living skills, that is, worse self-care and domestic or community skills, during pain than during a pain-free day. This contradicts the finding of another study, in which behavioral pain indicators of children with IDs after surgery were not related to daily living skills.

Exaggeration of Usual Symptoms of the Disability

The evidence for this behavioral pain indicator is derived from only 1 study with a 50% quality score; thus, cautiousness is recommended. “Exaggeration of usual symptoms of the disability” is reported to be an indication for acute period pain in women with Down syndrome. It remains unclear what is exactly meant.

Response to Analgesics

Similar to “exaggeration of usual symptoms of the disability,” the findings are derived from 1 study with a 50% quality score, so the evidence for analgesics as behavioral pain indicator in IDs is not very strong.

Parents report that response to analgesics is an indication for acute period pain in women with Down syndrome: symptoms of distress reduce and facial color returns after taking a paracetamol, and this effect lasts for 1 to 2 hours after the daily hours of action of paracetamol. Metabolic individual differences in people with IDs, however, may influence the actual response to analgesics.

Stereotyped Movements

The evidence provided by the study has a 100% quality score. Nonverbal children with IDs, probably severe to profound IDs, display stereotyped movements after surgery such as hands flapping, rubbing hands, or gnashing of teeth. It is hypothesized that these children, in contrast to verbal children with IDs, do not attempt to involve the people around them in their painful experience. In other words, stereotyped movement is not considered to be an intentional communication activity.

Discussion

Behavioral Pain Indicators in People With IDs

The goal of the present systematic review was to define behavioral pain indicators in people with IDs and to evaluate the methodological quality of studies on this topic. On the basis of 27 studies, we defined 14 categories of behavioral pain indicators. Categories that were reported in most studies were motor activity,
facial activity, social-emotional indicators, and nonverbal vocal expression. They consisted of different, sometimes contrasting, behavioral pain indicators, such as facial pallor or redness, and increased or decreased activity level.

A limitation of these categories is the diversity of behaviors and the difficulty discriminating between pain and other causes such as discomfort or anxiety. An increased frequency of chin raising combined with an increased intensity of chin raising and brow lowering during mild acute pain in adults with IDs\textsuperscript{23} was the most specific finding in the present review. The comparison between videotaped behavior during a vaccination condition and a sham condition\textsuperscript{31} increases the probability that the observed response during vaccination is caused by acute pain, and the participants function as their own control group. The mildness\textsuperscript{23} of the painful stimulus and the focus on only the face/upper torso,\textsuperscript{24} however, reduce the clinical relevance of the results. In that context, postoperative observations would be more informative concerning the detection of pain, although the likelihood of distress\textsuperscript{48} in postoperative settings confounds the discrimination between pain and other causes. When people with IDs appear to have painful medical conditions\textsuperscript{4} or when they react positively to analgesic medication,\textsuperscript{36} one might suspect that the behavioral disturbance is a reflection of pain. However, behavior management plans themselves may improve behavior instead of pain relief from medical conditions,\textsuperscript{4} and it remains unclear which cyclical behavior improves due to analgesic medication.\textsuperscript{36}

Although none of the studies in the current review included people with IDs at the same time as people without, the results suggest that people with IDs respond to pain or discomfort much the same way as the general population and as people with cognitive impairment,\textsuperscript{39} for example, with grimacing and guarding. Other behavioral pain indicators may be specific for the IDs population, such as self-injurious behavior, stereotyped movements, and what Kyrkou\textsuperscript{22} referred to as “exaggeration of usual symptoms of the disability.”

The instruments used in the studies of this review are comparable to those used in pain literature, such as the Facial Action Coding System to detect pain in people with dementia\textsuperscript{21} and observation scales for pain in people with dementia that cover aspects such as facial expressions, vocal reactions, posture, and emotional changes.\textsuperscript{53}

The studies focused especially on acute pain and often included children and adults in the same study. Evidence was based on opinion or observation. Most studies had a MMAT quality score of 50%. The 4 MMAT score criteria did not apply to all studies. As a result, some studies could never receive a 100% quality score. For example, criterion 4.4 did not apply to case series. Because of the variety of methodologies used in the studies, the MMAT was still the most appropriate tool for the current review.

**Factors That Influence Behavioral Pain Indicators in People With IDs**

To recognize pain expression in people with IDs, one should know not only the common behavioral pain indicators but also the factors that influence

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**Figure 2.** Factors in people with IDs that clearly do not influence behavioral pain indicators, clearly influence behavioral pain indicators, or in a complex manner influence behavioral pain indicators. See Discussion for more information.
the display of those indicators. The studies provide clear evidence for some factors and more complex evidence for others (see Fig 2). Clear evidence exists for 5 factors that do not influence pain expression in children with IDs. The first factor involves the duration of pain and the cause of pain, such as pain caused by injury or illness. The second factor involves medical and surgical characteristics, such as epilepsy, number of medications, and length of time in the recovery room. The third factor is psychopathology, such as mood disorders. The fourth factor involves daily living skills, such as getting dressed. The fifth factor is the living situation, that is, whether a child lives with its family or in a residential center.

Clear evidence also exists for 3 factors that do influence pain expression in children with IDs. The first influencing factor is pain intensity. As the intensity of pain rises, adaptive functioning decreases and the total number of behavioral pain indicators increases. The second influencing factor is verbal ability. Verbal children communicate their pain experience to others with, for example, complaining, whereas nonverbal children lack expressivity and show nonfunctional, stereotyped movements. The third influencing factor is a learned response to pain. Children could be influenced by previous experiences with pain and the pain responses of others. Clear evidence in adults with IDs exists for 1 factor that influences pain expression: the level of clinical dependence in cerebral palsy. For example, very dependent adults express pain when painful zones are touched or moved, whereas not very dependent adults protect painful zones (see Supplementary Table 3).

For 4 factors, the evidence about the influence on pain expression is more complex. The first complex factor is chronological age. Although children and adults with IDs differ in their expression of pain, with adults showing more individual differences, chronological age does not influence pain expression within these 2 age groups. The second complex factor is developmental age. Although developmental age is unrelated to pain expression in children with IDs, an indirect relationship was found in a group of both children and adults with IDs. A younger developmental age is related to less cognitively demanding coping styles such as helplessness and seeking social support, which, in turn, are related to a greater manifestation of behavioral pain indicators. The third complex factor is gender. Gender does not influence pain expression in children with IDs or in adults with IDs who are exposed to acute pain. However, it does influence adults with IDs who are exposed to uncomfortable sensory stimuli such as deep pressure and pin prick. In the latter case, females display more facial activity than men. The fourth complex factor is level of IDs. In adults with IDs who are exposed to acute pain, having a more severe level of IDs resulted in a display of more behavioral pain indicators, but the level of IDs was unrelated to the pain intensity observed on the basis of facial activity. This could mean that facial activity is sensitive to acute pain in adults with all levels of IDs, and that the total number of behavioral pain indicators displayed depends on the level of IDs.

**Conclusion**

As far as we know, this is the first systematic review about behavioral pain indicators in people with IDs. Fourteen categories of behavioral pain indicators were defined that can be useful to detect pain. Sometimes, however, the categories consisted of contrasting behavioral pain indicators, and specific factors may influence the pain expression in people with IDs. Moreover, it is often unclear which part of the observed behavior is caused by pain and which by other factors such as stress or fear. Because of these complicating aspects and because most studies had an MMAT quality score of 50%, one has to interpret the present findings with caution.

**Recommendations for Clinical Practice**

Caregivers should be aware of the various behavioral pain indicators, listed in Table 2, and the factors that influence them. The routine use of pain observation checklists that are validated for people with IDs is recommended, such as the Non-Communicating Children Pain Checklist Postoperative Version and the Non-Communicating Adult Pain Checklist. The advantage of standardized instruments is that they can be used by observers who are not familiar with the individual with IDs. Virtually all instruments used in the studies of this review have moderate or good psychometric properties and are described as suitable for clinical use. Scales for pain intensity such as visual analog scales or numeric rating scales correlate with observation scales in people with IDs and can thus be combined with observation scales for a more complete approach in pain diagnostics. Besides general categories, individual-specific indicators are useful to detect pain in people with IDs, and the identification of such indicators could possibly reduce problem behavior during pain. The Individualized Numeric Rating Scale could be used to make a profile for each individual with IDs, including baseline behavior, observed responses to a known pain source, and current behavior. A medical examination in people with IDs who show self-injurious behavior may reveal painful medical conditions that first need to be treated, for example, duodenal ulcers and oral abscesses.

**Recommendations for Further Research**

The complexity of the IDs population should not hinder researchers in performing more studies on behavioral pain indicators, as knowledge about this topic is still in its infancy. The following studies in the IDs population are needed: studies that are experimentally controlled, studies that use naturally occurring acute painful procedures such as acupuncture, studies that address chronic pain, studies that involve further psychometric evaluation of existing...
pain observation scales instead of developing new scales, and studies that focus on a better differentiation between pain and mental states such as anxiety. Behavioral patterns specific for pain in people with IDs are more or less unknown. More knowledge and easy access to this knowledge for the clinical field will provide caregivers guidance in detecting pain and will stimulate research on interventions for pain treatment in the IDs population. This could consequently result in an improved quality of life in people with IDs who suffer from undiagnosed pain.

Supplementary Data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.jpain.2013.04.016.

References


