

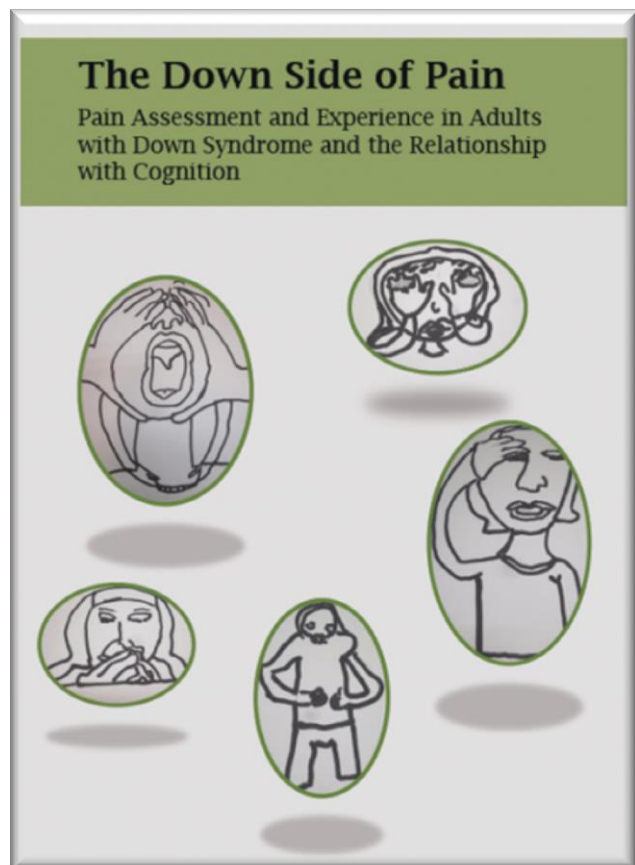
Pain experience in adults with Down syndrome

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The Committee for Science & Society of the Trisomy 21 Research Society (T21RS) regularly addresses issues raised by parents and Down syndrome associations through summarizing the state-of-the-art knowledge from a scientific perspective. The Committee for Science & Society* is strongly committed to introducing scientific research and explaining recent findings in an understandable way. Today, the third edition of the T21RS Science & Society Bulletin: key issues on pain experience in Down syndrome.

KEY QUESTIONS ABOUT PAIN IN DOWN SYNDROME

- What is already known?
- Is pain experience related to mental functions?
- How to ask about pain experience?
- How to recognize behavioural pain signals?
- What is a practical take-home message?



Thesis, Nanda de Knegt, 22-09-2015, VU University, Amsterdam (pain expressions are drawn by a participant of the PhD study).

What is already known?

People with Down syndrome have an increased risk for physical conditions that may be painful, such as middle ear infections, skin problems, hip and knee instability, and foot deformities. The risk could increase even further due to a rising life expectancy of the Down syndrome population. This is for example reflected in an early onset of arthritis. It can be concluded that awareness of the presence of painful and discomforting conditions is especially needed in adults with Down syndrome.

The pain experience in Down syndrome is however unclear, because impairments in mental functions and communication could hamper self-report of pain and people with Down syndrome may have a tendency to express medical problems with problematic behaviour instead of complaining about pain. The brain areas that are responsible for pain processing may be different in people with Down syndrome, which could result in an increased or decreased pain experience.



According to the most recent overview of scientific literature on the topic, people with Down syndrome could be actually more sensitive to pain but with a delayed pain expression. This delayed pain response may be caused by a disturbed transmission of pain signals, for example from the body to the brain or within the brain. The higher pain sensitivity may be caused by a magnified nerve signal of pain and/or an inefficient inhibition of pain. However, scientific evidence for a higher pain experience in Down syndrome is limited to only two findings: 1) a longer persisting pain response after medical procedures in babies with Down syndrome compared to a control group and 2) a lower threshold for pain due to heat in Down syndrome compared to a control group after correction for reaction time. In addition, research results do not indicate differences between children with and without Down syndrome in the requirements or the metabolism of pain medication after surgery.

Pain assessment is difficult in people with intellectual disabilities. Caregivers report that recognizing and treating pain in adults with intellectual disabilities is complex. This is alarming, because under-treatment of pain in people with intellectual disabilities has been reported in the scientific literature. In a Dutch study, 66% of the parents stated that their child with Down syndrome is less sensitive to pain than the siblings. However, caregivers of people with intellectual disabilities were already warned by a researcher in 1994 that they “may miss signs of illness or injury because they are looking for the more obvious pain signals”. Pain behaviour specific for Down syndrome is unknown. Two research groups have examined the use of scales to self-report pain in people with Down syndrome, showing that an enlarged body map is needed to point to the painful location and that a facial scale may be better understood than a coloured scale to rate pain intensity expressed in pictures of painful situations.

Is pain experience related to mental functions?

Pain assessment is important, because chronic pain could have a negative influence on emotional well-being, quality of life, and mental functions that are needed in daily life. Examples of the mental functions that are vulnerable to pain are concentration, memory, and abstract thinking. However, a relationship between self-reported pain experience (average of ratings in rest and during a series of movements such as walking) and mental functioning (tests for memory and complex functions such as planning) was not found in 232 adults with Down syndrome. Although structural differences and atypical patterns of brain activation in adults with Down syndrome arise the question whether the same brain areas for pain experience and cognitive functioning are involved and activated in Down syndrome as in the general population, the absence of a relationship could also be explained by a low pain intensity level during the test session and a small number of participants in the final analyses (who reported pain, understood the pain scales, and performed all cognitive tests).

How to ask about pain experience?

Difficulties in pain assessment of people with intellectual disabilities include: 1) a tendency to answer “yes” to every closed question and to repeat the final option in questions with multiple answers, and 2) impaired understanding of abstract concepts such as time (for example: “How long have you had the pain?”). In addition, pain assessment in people with Down syndrome is hampered by deficits in expressive language abilities. As a result, self-report of pain experience by people with intellectual disabilities is unfortunately rarely used in research. Tools for self-reporting of pain may provide individuals with intellectual disabilities with a sense of self-determination and a feeling of greater control over their lived experience. Adults with Down syndrome comprehended a facial scale (75%) better than a numeric scale (43%) and almost 80% comprehended at least one of these scales. When the comprehension of the numeric scale was assessed thoroughly, by not only asking which numbers represent the least pain (0 or 1) and the most pain (9 or 10) but also asking questions about the magnitude of numbers (“Which is larger: 2 or 8?” and “Which is larger: 6 or 4?”), then fewer participants passed the comprehension test. Facial pictograms were comprehended and preferred as much as drawn faces. Half of the participants understood a series of pictograms for the sensory-discriminative pain aspect (burning, stinging, throbbing, and pressing).

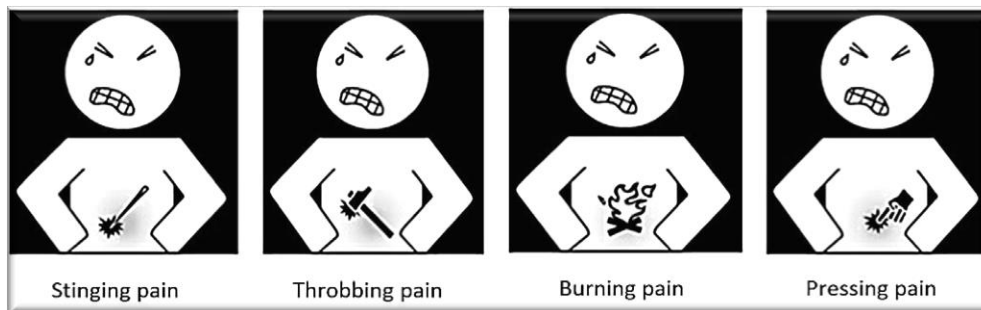
How to recognize behavioural pain signals?

An initial step in the Dutch national guideline ‘Signalling pain in people with intellectual disabilities’ is the detection by caregivers that something is wrong. Because not all adults with Down syndrome are able to understand tools for self-report of pain, behavioural pain signals are important. Although little is known about pain behaviour specific for Down syndrome, an overview of scientific literature showed that people with intellectual disabilities show behavioural pain indicators that correspond to universal categories (such as facial expression, movements/posture, and physiological indicators) but also include less obvious expressions (such as aggression, agitation, depression, self-mutilation, and stereotype movements). ‘Problem behaviour’ could be an expression of pain, which is important regarding the large amount of antipsychotics that are prescribed for people with intellectual disabilities. Knowledge about pain expression in an individual is crucial, because expressions differ within universal categories and observational checklists may miss less obvious expressions.



What is a practical take-home message?

- Remain vigilant for painful physical conditions in adults with Down syndrome.
- Be aware that a possible delayed pain response, less obvious pain behaviour, and difficulties with self-report could lead to an incorrect assumption that people with Down syndrome have a higher pain threshold or lower pain sensitivity.
- Stimulate adults with Down syndrome to talk about pain (using open, concrete questions).
- The possibility for self-reporting pain experience should be examined to enhance a sense of self-regulation
- Family members or the 'pain team' of an organization could investigate which pain scale is best understood by a person with Down syndrome.
- Input from the family is crucial for information about the individually characteristic behavioural expressions of pain.
- Although it probably will remain difficult for people with Down syndrome to understand the various types of pain (e.g., stinging), it is expected that practicing the use of pictograms to talk about pain experience will increase the advantage of these pictograms.
- Remain vigilant to a change in mental functions when people with Down syndrome are in pain. Examples are attention, memory, mental flexibility, and the ability to regulate the own behaviour.



* The Committee for Science & Society consists of prof. dr. Peter Paul De Deyn (chairman, Belgium), Juan Fortea (Spain), Sebastián Videla (Spain), Cindy Lemere (USA) and Alain Dekker (The Netherlands).



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Bibliography: 5 key references (other references can be found in the thesis of Nanda de Knegt)

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The thesis of Nanda de Knegt can be requested without charge via nc.de.knegt@vu.nl Questions about the current bulletin could also be asked via this e-mail address.