Pijn experience en cognition in Williams, Prader-Willi en Fragiele-X syndromes

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The rationale is to provide practical tools for pain assessment (for both the professional and parents) and to understand the pain experience in people with WS, PWS, and FXS for developing/adapting pain management.The research questions are:1) What...

Ethical review	Approved WMO
Status	Recruitment stopped
Health condition type	Chromosomal abnormalities, gene alterations and gene variants
Study type	Observational non invasive

Summary

ID

NL-OMON47146

Source
ToetsingOnline

Brief title
Pijn bij Williams, Prader-Willi en Fragiele-X syndromen

Condition

- Chromosomal abnormalities, gene alterations and gene variants

Synonym
chromosome 15 en X chromosome, Prader-Willi syndrome en Fragiele-X syndromes; abnormality in chromosome 7, Williams syndrome

Research involving
Human

Sponsors and support

Primary sponsor: Vrije Universiteit
Source(s) of monetary or material Support: Ministerie van OC&W,Vrije Universiteit (eigen bijdrage);Stichting Sociaal-Pedagogische Zorg;Jan Jongmans Fonds en Cornelia Stichting

Intervention

Keyword: Cognition, Intellectual disabilities, Pain assessment, Pain experience

Outcome measures

Primary outcome

The primary parameters are: 1) the difference in the pain characteristics between clinical groups and between clinical groups and control groups; and 2) the relationship between pain experience and cognitive function in the clinical groups.

Secondary outcome

Not applicable.

Study description

Background summary

In people with Williams syndrome (WS), Prader-Willi syndrome (PWS) and Fragile X syndrome (FXS), there is evidence of painful physical conditions and indications for another pain perception and sensory sensitivity. Our research in people with dementia and adults with Down syndrome has shown that the specific characteristics of brain areas and white matter tracts involved in pain processing may result in a different perception of pain and that it is important to assess pain reliably. Additionally, the increased life expectancy in the population of people with intellectual disabilities has resulted in a greater risk of painful medical conditions, allowing more clients will be with pain who might otherwise experience pain and / or pain may indicate less well. Furthermore, pain is negatively related to physical inactivity: in the general population there are those without dementia and with evidence that physical inactivity can result in pain and that pain can lead to physical inactivity. This is worrisome because it has been found that older people with intellectual disabilities are very physically inactive. Evidence for physical inactivity has also been found in adolescents and adults with WS, PWS and FXS. In addition to physical inactivity has hurt also negatively related to mood, behavior, sleep pattern, adaptive functioning and quality of life. Caregivers of adults with intellectual disabilities have observed that pain can lead to For example anxiety / stress, anxiety, depression, self harm and problem behaviour. This

could be the only expression of pain due to the sometimes difficult self-report. Possible examples of behavioral problems related to pain his temper and beat others. The areas of adaptive functioning receding in pain in children with intellectual disability are communication, social skills, motor skills and daily activities. Finally, pain associated with poorer cognitive function, as found in chronic pain patients regarding decisions, attention, memory, working memory, word fluency and cognitive flexibility. These functions are essential to daily functioning.

Abnormal pain behavior (such as self-injury), difficulty with self-reporting pain (resulting in vague descriptions), or a high pain threshold in some individuals with intellectual disabilities can make it difficult for caregivers and professionals to notice pain in time. Caregivers of adults with intellectual disabilities have reported that pain diagnostics and treatment are complex and unclear. The possibilities of self-reports of pain by people with intellectual disabilities must be carefully investigated, because the people themselves deliver the most important information (self-report is the gold standard for measuring pain) and it stimulates autonomy to ask about their experience. In subjects who have difficulties communicating pain it is valuable to examine which self-reporting scale (numbers, faces, or icons) is best understood. In subjects who can communicate pain it is important to examine the pain experience, so that this knowledge can be used to notice and treat pain. At present there is a lack of knowledge about the pain experienced by people with WS, PWS, and FXS. The purpose of this study is to increase this knowledge and to be of direct use for the individual.

To obtain more insight into pain experience, it is valuable to examine the relationship with cognitive function (mental functions such as memory, attention, and planning). Many brain areas that proces pain also have a cognitive function and a negative relationship has been found between pain and cognition. It is therefore relevant to examine whether a worse cognitive functioning is an indication for pain. It is further relevant to examine in what extent the self-reported pain correspond to observed pain behaviour and the presence of potentional painful conditions. When describing potentially painful conditions, attention should also be paid to dental conditions. The oral health of people with intellectual disabilities is in general concerning, such as a higher prevalence and greater severity of periodontal disease. Possible causes include communication problems, no cooperation, and motor or cognitive limitations. Poor oral health can lead to pain. Acute pain from an abscess in the mouth can result in people with intellectual and communicative limitations in aggressive behaviour. Poor oral health also results in a risk of diseases such as pneumonia.

Study objective

The rationale is to provide practical tools for pain assessment (for both the professional and parents) and to understand the pain experience in people with

WS, PWS, and FXS for developing/adapting pain management.

The research questions are:
1) What are the characteristics of pain (presence, perception, behaviour, sensory sensitivity) in WS, PWS, and FXS, compared with two control groups from the general population without diagnoses of genetic abnormalities (matched for chronological and mental age)?
2) Is there a relationship between pain perception and cognitive function in people with this syndrome?

Study design

Observational study (case-control). The research provides the most complete and careful approach as possible by combining information from self-report (at rest and after exercise), report of caregivers, files form physician and dentist, observation of pain behaviour (by researcher and caregivers), and sensory and cognitive test results.
(See Chapter 8 of the study).

Study burden and risks

Risks are absent (no painful stimuli will be administered) and the burden is reduced by preventing exhaustion (taking breaks and using several testing sessions, if necessary). A previous study of pain experience and cognition in adults with Down syndrome and the pilot study of the current project shows that a test session of three hours is not too strenuous. During the test session, close attention will be paid to fatigue and resistance of participants and participants will be encouraged.

Contacts

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Trial sites

Listed location countries

Netherlands

Eligibility criteria

Age
Adolescents (12-15 years)
Adolescents (16-17 years)
Adults (18-64 years)
Children (2-11 years)
Elderly (65 years and older)

Inclusion criteria

Inclusion criteria of the clinical groups are:
- Wiliams, Prader-Willi, or Fragile-X syndrome according to genetic assessment
- Adult age (18 years and older)
- Capable of speaking;Inclusion criteria of the control groups are:
- Control group matched on calendar age: adults (18 years and older)
- Control group matched on mental age: children 2-17 years
- Participants in both control groups are capable to speak Dutch

Exclusion criteria

Exclusion criteria of the clinical groups are:
- Severe level of intellectual disability
- Comorbid psychiatric disorder due to which the test session is not possible
- Visual or auditory impairments due to which the test session is not possible
- Advanced stage of dementia due to which the test session is not possible;Exclusion criteria of the control groups are:
- Indication of 'special education': attending such education in present or past
- Diagnosis of a genetic abnormality
- Diagnosis of a neurological condition
- Diagnosis of a disorder in speaking or understanding language
- Diagnosis of emotional disorders (such as depression, bipolar disorder, anxiety disorder), attention deficit or behavioural disorder (such as ADD, ADHD, ODD, CD) autism or autism spectrum disorder (such as PDD-NOS), or mental disorders (such as borderline, schizophrenia, OCD, psychoses)

- Visual or auditory impairments due to which the test session is not possible
- Use of antipsychotic, anticonvulsant, or antidepressive medication
- Use of a wheel chair (due to movements during the test assessment)

Study design

Design

Study type:	Observational non invasive
Intervention model:	Other
Allocation:	Non-randomized controlled trial
Masking:	Open (masking not used)
Control:	Active
Primary purpose:	Diagnostic

Recruitment

NL

Recruitment status:	Recruitment stopped
Start date (anticipated):	24-12-2016
Enrollment:	200
Type:	Actual

Ethics review

Approved WMO Date:	15-12-2016
Application type:	First submission
Review commission:	METC Amsterdam UMC
Approved WMO Date:	26-04-2017
Application type:	Amendment
Review commission:	METC Amsterdam UMC
Approved WMO Date:	15-01-2018
Application type:	Amendment

Review commission:	METC Amsterdam UMC
Approved WMO Date:	08-02-2019
Application type:	Amendment
Review commission:	METC Amsterdam UMC

Study registrations

Followed up by the following (possibly more current) registration

No registrations found.

Other (possibly less up-to-date) registrations in this register

No registrations found.

In other registers

Register	ID
CCMO	NL54788.029.16