Physical conditions and challenging behaviour in people with intellectual disability: a systematic review

C. F. de Winter,¹ A. A. C. Jansen² & H. M. Evenhuis³

¹ Reinaerde, Organisation for People with Intellectual Disability, Den Dolder, the Netherlands
² Centre for Consultation and Expertise, Utrecht, the Netherlands
³ Erasmus MC, General Practice, Intellectual Disability Medicine, Rotterdam, the Netherlands

Abstract

Background Challenging behaviour is a major problem among people with intellectual disabilities. Physical factors may be an important cause. The aim of the present systematic review was to determine the physical conditions associated with challenging behaviour.

Methods A literature search was conducted in PubMed and the Cochrane systematic review database for empirical studies published between 1990 and 2008. The quality of all the studies that met the inclusion criteria was assessed using the SIGN-50 methodology checklists.

Results The search identified 45 studies, which looked at general medical conditions, motor impairment, epilepsy, sensory impairment, gastrointestinal disease, sleep disorders, dementia and others. There were four high-quality observational studies, seven well-conducted observational studies, 21 observational studies of low methodological quality and 13 non-analytical studies. There were significant and independent associations between challenging behaviours and urinary incontinence, pain related to cerebral palsy and chronic sleep problems, and between self-injurious behaviour and visual impairment. No association was found with hearing impairment, bowel incontinence, mobility impairment or epilepsy. Many other physical conditions were not addressed at all.

Conclusion Medical conditions can play a role in challenging behaviour, and this should be evaluated in the clinical setting. So far, the level of evidence is generally low, and longitudinal studies are completely lacking. We recommend a systematic approach to research examining the role of physical conditions in challenging behaviour, the ultimate aim being to establish a basis for the development of clinical guidelines.

Keywords aggression, challenging behaviour, intellectual disability, medical conditions, self-injurious behaviour, systematic review

Introduction

Challenging behaviour is a major problem in people with intellectual disabilities (ID). The estimated prevalence of these severe behaviour problems in large population studies is between 10% and 15% (Harris 1993; Sigafos et al. 1994; Emerson et al. 2001a; Holden & Gitlesen 2006; Jones et al. 2008).
The most common forms of challenging behaviour are aggression (7%), destructive behaviour (4–5%) and self-injury (4%; Emerson et al. 2001a). Furthermore, the most demanding forms of these behaviours tend to persist throughout life (Emerson et al. 2001b; Murphy et al. 2005; Totsika et al. 2008). Challenging behaviours can have serious disadvantages for the children and adults in question, such as interference with development, quality of life, social participation (Holden & Gitlesen 2006), physical injury and even death (Noel & Clarke 1982; Jan et al. 1994; Carllock et al. 1997; Nissen & Haveman 1997). People displaying aberrant behaviours also tend to be over medicated (Matson et al. 2000; Stolker et al. 2008). Family and staff can find it very stressful and emotionally difficult to deal with people with challenging behaviour (Hastings & Brown 2002).

The causal and maintaining mechanisms underlying challenging behaviour are multifactorial (Applegate et al. 1999; Schroeder et al. 2001; McClintock et al. 2003; Matson & Boisjoli 2007; Tenneij & Koot 2008). Although functional analysis and treatment have focused mainly on behavioural aspects (Hassiotis & Hall 2008), physical factors also play an important role in different forms of challenging behaviour (self-injury, aggression, stereotypes, pica and rumination) (Applegate et al. 1999; Matson & Boisjoli 2007). Bosch et al. (1997) performed an exploratory study in people with self-injurious behaviour (SIB) and ID. In 28% of the patients, they found previously undiagnosed medical conditions that could be expected to cause pain or discomfort. Conditions that may cause pain (and may go unnoticed) are inflammatory or infectious diseases, motor impairment, pulmonary or cardiac disease, gastrointestinal conditions, malignancies, lacerations or fractures, headache, ear, nose, throat or eye diseases (Bosch et al. 1997; Ryan & Sunada 1997; van Schrojenstein Lantman-de Valk & Walsh 2008). People with ID and SIB have intact pain sensation (Symons et al. 2009) and SIB can target the site of the pain (Breau et al. 2003). Pain may very well be a cause of SIB (Symons et al. 2009; Oliver & Richards 2010), and one can imagine that this is also the case for other types of challenging behaviour, especially if people are unable to express their complaints otherwise (Kastner et al. 2001).

It has been proposed that challenging behaviours should be assessed on multidisciplinary lines and that medical conditions should be taken into account (Loschen & Osman 1992; Kwok & Cheung 2007; The National Association for the Dually Diagnosed 2007). The aim of this systematic review is to determine which physical conditions are associated with challenging behaviour.

**Methods**

**Selection criteria for studies covered by this review**

**Types of studies**

We considered relevant empirical/observational studies published between January 1990 and July 2008 with a minimum sample size of five participants. The papers were written in English, Dutch or German.

**Types of participants**

Participants were children and adults with ID. All levels of ID were included.

**Types of exposure**

All physical medical conditions were included. Medication side effects and substance abuse were excluded. We excluded studies of specific syndrome phenotypes, although syndromes may include both physical conditions and specific behavioural problems. Behavioural problems are widely considered to be related to the syndrome (and psychiatric profile) rather than to physical co-morbidity, and possible causal relationships are not studied. We do not intend to describe behavioural phenotypes.

**Types of outcome**

We defined challenging behaviour using the description of problem behaviours in ‘Diagnostic criteria for psychiatric disorders for use with adults with learning disabilities (DC-LD)’, axis III, level D: general diagnostic criteria for problem behaviours, verbally aggressive behaviour, physically aggressive behaviour, destructive behaviour, self-injurious behaviour, sexually inappropriate behaviour, oppositional behaviour, demanding behaviour, wandering
behaviour, mixed problem behaviour, other problem behaviours and mixed other problem behaviours. We also included problematic feeding disorders: food rumination/regurgitation disorder and pica, DC-LD axis III, level B (Royal College of Psychiatrists 2001).

Search methods for identification of studies

Electronic searches

We searched Medline/PubMed and the Cochrane Database of Systematic Reviews. Research terms were selected to search very broadly on ID, all forms of problem behaviours and all possible medical conditions that might be associated with behavioural problems (based on known literature and clinical practice). Terms were searched for as MeSH terms when indicated, and otherwise they were searched for in all fields (title, abstract, keywords).

In a first search, research terms for intellectual disability were combined with terms for problem behaviour. In a second search, terms for intellectual disability were combined with terms for medical conditions and ‘behaviour’.

Terms for intellectual disability were: ‘Mental Retardation [MeSH]’ OR ‘Intellectual Disability’ OR ‘Developmental Disability’ OR ‘Mental Handicap’ OR ‘Multiple Handicap’ OR ‘Intellectual Impairment’ OR ‘Communicative Impairment’ OR ‘Learning Disability’ OR ‘Neurodevelopmental Disability’ OR ‘Cognitive Impairment’.


Searching other resources

We scrutinised the bibliographies of articles that seemed relevant.

Data collection and analysis

Selection of studies

One reviewer (C. F. dW.) screened the titles and abstracts from the search. The full texts of papers that appeared relevant were retrieved. Four articles were available as epub ahead of print at the time of the search. The actual publication dates were outside our inclusion period, but they were included in this review. A total of 45 articles fulfilled our inclusion criteria and they were included in the present review.

Data extraction and management

Two reviewers (C. F. dW. and H. M. E.) then analysed the study characteristics and methodological quality/level of evidence of the articles independently. Where there were gaps in the available information, we attempted to contact the authors. When the reviewers judged the articles differently, they discussed the results in order to reach agreement. A third reviewer was on hand to make a final decision if agreement could not be reached. This was not necessary as full agreement was reached about the study characteristics and the level of evidence of all articles.

A note was made about the following study characteristics for all articles:

- Study design.
- Participants: number of participants, age of the participants, sex, level of ID and study population...
• Exposure: the medical conditions discussed, evaluation method and reliability (Table 1).
• Outcome: kind of problem behaviour, and assessment of behaviour and reliability (Table 1).
• Results of the study and relevant conclusion for the present review.

Assessment of the level of evidence

At present, there is no consensus about a tool for the assessment of quality in observational studies (Sanderson et al. 2007). For case–control studies and for cohort studies (SIGN 2008), we used the SIGN-50 methodology checklists as they are designed for observational studies and comprise most of the items indicated by the STROBE statement for the publication of observational studies (von Elm et al. 2007). Although SIGN-50 was originally designed as a basis for guideline development, we used it as a quality tool for the present systematic review. We used the SIGN methodology checklist for cohort studies and adapted this for cross-sectional studies. The SIGN-50 methodology checklists include items covering internal validity (study question, selection of participants, assessment, confounding and statistical analysis), the overall assessment of the study and the description of the study. We checked whether all items indicated by the STROBE guidelines for reporting on observational studies (von Elm et al. 2007) were adequately addressed by the SIGN checklists. We also looked at whether a power analysis had been conducted, because the STROBE guidelines require reports of observational studies to explain how study size is determined (von Elm et al. 2007).

Accordingly, the studies were rated for level of evidence (LE) using the SIGN criteria (SIGN 2008; Table 2). The SIGN criteria do not mention cross-sectional studies and we therefore considered cross-sectional studies without any statistical analysis to be non-analytical studies (LE 3). We allocated cross-sectional studies with statistical analysis to the same category as case–control and cohort studies (LE 2).

Results

The 45 articles identified in the search were allocated to eight different categories according to the type of exposure: physical conditions in general, motor disorders, sensory impairment, epilepsy, gastrointestinal disease, sleep disorders, dementia and others. There were many categories about which no articles were found, examples being menopause, cardiac and pulmonary disease, infectious disease and malignancies. Most articles addressed our review area as a secondary issue. In these cases, our quality assessment was confined to the questions related to the review area. The results of the review of study characteristics, study results and level of evidence have been described by category and they can be found in Tables 3–10. All studies included both males and females, with the exception of one...
male-only (Day 1994) and one where female-only study (Taylor et al. 1993). Many of the study groups were very heterogeneous, including participants with both known and unknown aetiology. Furthermore, many studies had a cross-sectional design but they were also retrospective, being based on file data. This area is addressed in Tables 3–10 under the headings Exposure and Outcome measures.

Physical conditions in general

Our search identified 11 cross-sectional studies looking at general medical conditions and challenging behaviour (Table 3). The quality of the studies varied: there were five non-analytical studies, three analytical studies with a high risk of bias and three high-quality studies (Table 3).

The three high-quality studies all looked at the same study population. A study of problem behaviour in general found a significant correlation with visual impairment, urinary incontinence and not having a severe physical disability. There was no association with hearing impairment, bowel incontinence, impaired mobility and epilepsy (Jones et al. 2008). In the same study population, there was a significant correlation between SIB and visual impairment. There was no association with hearing impairment, bowel and urinary incontinence, impaired mobility and epilepsy (Cooper et al. 2009a). Urinary incontinence was significantly and independently associated with aggressive behaviour in adults, whereas no significant correlation was found for vision impairment, hearing impairment, bowel incontinence, impaired mobility and epilepsy (Cooper et al. 2009b). In studies with a higher risk of bias, Collacott et al. (1998) found an association between SIB and hearing impairment and impaired mobility, but not with epilepsy and visual impairment. Deb et al. (2001) found the same prevalence of behavioural disorders in people with or without physical disabilities or illness.

Low-quality studies suggest that previously undiagnosed and acute medical conditions are associated with severe problem behaviours. The disease categories are gastrointestinal (lactose intolerance, constipation, colon carcinoma), neurological (epilepsy, Parkinson’s, sleep apnea, migraine, stroke, encephalitis, hypothalamic hamartoma), urogenital (urinary tract infection, renal insufficiency, pelvic inflammatory disease, gynaecological malignancies, prostatitis), ear–nose–throat (otitis media, sinus problems, dental disease), cardiological (valve disease, heart failure), trauma (lacerations, broken bones, head trauma), eye diseases (dry eyes, cataracts), endocrine (diabetes mellitus, thyroid disease), pulmonary (chronic obstructive pulmonary disease, lung cancer), dermatological (eczema, cellulites), musculo-skeletal (arthritis, scoliosis), haematological (anaemia) and others (dehydration; Peine et al. 1995; Bosch et al. 1997; Ryan & Sunada 1997; Kastner et al. 2001). Davidson et al. (1994) found no differences in medical determinants between people with aggressive behaviour compared to people with other behavioural problems.

In conclusion, urinary incontinence is associated with challenging behaviour (high level of evidence). Visual impairment is associated with SIB, but not with aggressive behaviour (high level of evidence). People with severe physical disabilities have a lower risk of problem behaviours (moderate level of evidence). Previously undiagnosed medical conditions and acute medical conditions are frequently noted in people exhibiting these behaviours. Some of these conditions cause acute or chronic pain (very low level of evidence).

Motor disorders

Two cross-sectional studies looking at cerebral palsy and one at non-ambulatory persons were identified. One of them was a high-quality study, one was an analytical study with a high risk of bias and the third was a non-analytical study (Table 4).

In the high-quality study, which examined children with cerebral palsy, almost 25% had significant behaviour problems. Children with more severe functional limitations had fewer behaviour problems, while children with severe pain had more problems. Sensory impairments and seizures were not significantly correlated with behaviour in children with cerebral palsy (Parkes et al. 2008). Blacher & McIntyre’s (2006) study suggested that young adults with cerebral palsy and ID had fewer internalising and externalising behaviour problems and were less aggressive than young adults with ID only and equal SIB and sexual problems, but this remains insufficiently proven. Kobe et al. (1994) noted many problem behaviours in non-ambulatory
Table 3  Characteristics and level of evidence of studies on general medical conditions

<table>
<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>n</th>
<th>Age</th>
<th>Level ID</th>
<th>Study population</th>
<th>Exposure measure</th>
<th>Outcome measure</th>
<th>Analysis &amp; results</th>
<th>Level of evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cooper et al. (2009a)</td>
<td>Cross-sectional</td>
<td>1023</td>
<td>Adults</td>
<td>All</td>
<td>Representative</td>
<td>**21st Century Health Check</td>
<td>**SIB by DC-LD</td>
<td>Multivariate logistic regression: OR = 1.939, P = 0.041 for visual impairment and SIB</td>
<td>2++</td>
</tr>
<tr>
<td>Cooper et al. (2009b)</td>
<td>Cross-sectional</td>
<td>1023</td>
<td>Adults</td>
<td>All</td>
<td>Representative</td>
<td>**21st Century Health Check</td>
<td>**Aggression by DC-LD</td>
<td>Multivariate logistic regression: OR = 1.995, P = 0.007 for urinary incontinence and aggression</td>
<td>2++</td>
</tr>
<tr>
<td>Jones et al. (2008)</td>
<td>Cross-sectional</td>
<td>1023</td>
<td>Adults</td>
<td>All</td>
<td>Representative</td>
<td>**21st Century Health Check</td>
<td>**Problem behaviour by DC-LD</td>
<td>Multivariate logistic regression: visual impairment OR = 1.460, P = 0.033, urinary incontinence OR = 2.053, P = 0.000, severe physical disabilities OR = 0.179, P = 0.000</td>
<td>2++</td>
</tr>
<tr>
<td>Deb et al. (2001)</td>
<td>Cross-sectional</td>
<td>101</td>
<td>Adults</td>
<td>All</td>
<td>Representative</td>
<td>**Purpose-designed questionnaire (physical disabilities or illness)</td>
<td>**Psychiatric interview and Disability Assessment Scale</td>
<td>No significant difference in behaviour disorders in people with and without physical impairment or illness</td>
<td>2-</td>
</tr>
<tr>
<td>Kastner et al. (2001)</td>
<td>Cross-sectional</td>
<td>209</td>
<td>Adults</td>
<td>All</td>
<td>Referred</td>
<td>**Medical files (retrospectively)</td>
<td>**Behaviour rating scale: SIB, aggressive, disruptive, in appropriate habits, others</td>
<td>12% undiagnosed condition†</td>
<td>3</td>
</tr>
<tr>
<td>Collacott et al. (1998)</td>
<td>Cross-sectional</td>
<td>2101</td>
<td>Adults</td>
<td>All</td>
<td>Representative</td>
<td>**Not described</td>
<td>**SIB by interview and Disability Assessment Schedule</td>
<td>Backward stepwise logistic regression analysis: hearing impairment and immobility P = 0.0001</td>
<td>2-</td>
</tr>
<tr>
<td>Bosch et al. (1997)</td>
<td>Case series</td>
<td>25</td>
<td>Children &amp; adults</td>
<td>Moderate to profound</td>
<td>Referred</td>
<td>**Medical files (retrospectively)</td>
<td>**SIB</td>
<td>28% undiagnosed condition that may have caused pain or discomfort†</td>
<td>3</td>
</tr>
<tr>
<td>Study</td>
<td>Design</td>
<td>n</td>
<td>Age</td>
<td>Level ID</td>
<td>Study population</td>
<td>Exposure measure</td>
<td>Outcome measure</td>
<td>Analysis &amp; results</td>
<td>Level of evidence</td>
</tr>
<tr>
<td>------------------------------</td>
<td>----------------</td>
<td>-----</td>
<td>---------</td>
<td>----------</td>
<td>------------------</td>
<td>---------------------------------------------------------------------------------</td>
<td>--------------------------------------------------------------------------------</td>
<td>---------------------------------------------------------------------------------</td>
<td>--------------------</td>
</tr>
<tr>
<td>Ryan &amp; Sunada (1997)</td>
<td>Cross-sectional</td>
<td>1135</td>
<td>Adults</td>
<td>Moderate to severe</td>
<td>Referred</td>
<td>***Medical examination and additional test protocols (medical files, retrospectively)</td>
<td>Not described</td>
<td>75% previously undiagnosed or undertreated medical condition†</td>
<td>3</td>
</tr>
<tr>
<td>Peine et al. (1995)</td>
<td>Case series</td>
<td>10</td>
<td>≥45 years</td>
<td>All</td>
<td>Residence</td>
<td>Medical files</td>
<td>Staff records on SIB, aggregation, agitation, noncompliance, self-stimulation</td>
<td>Challenging behaviour may coincide with acute medical conditions†</td>
<td>3</td>
</tr>
<tr>
<td>Davidson et al. (1994)</td>
<td>Cross-sectional</td>
<td>199</td>
<td>Children &amp; adults</td>
<td>All</td>
<td>Referred</td>
<td>Medical files</td>
<td>Medical files</td>
<td>files and intake chart</td>
<td>3</td>
</tr>
<tr>
<td>Hyman et al. (1990)</td>
<td>Cross-sectional</td>
<td>97</td>
<td>Children</td>
<td>Not described</td>
<td>Referred</td>
<td>Medical files (retrospectively): cerebral palsy, sensory impairments, otitis media, seizures</td>
<td>SIB</td>
<td>Comparison to published data. Co-occurrence of SIB and visual impairment and a history of infantile spasms†</td>
<td>3</td>
</tr>
</tbody>
</table>

Exposure and Outcome measure *, **, ***: see Table 1.
Level of evidence 2++, 2+, 3: see Table 2.
All = mild to profound ID.
Representative = study population is representative for ID population.
Referred = referred patients to a multidisciplinary team for people with ID and/or challenging behaviour.
Residence = study population is derived from a residence for people with ID.
ID, intellectual disabilities; SIB, self-injurious behaviour; DC-LD, diagnostic criteria for psychiatric disorders for use with adults with learning disabilities.
† No statistical analysis.
<table>
<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>n</th>
<th>Age Level</th>
<th>Study population</th>
<th>Exposure measure</th>
<th>Outcome measure</th>
<th>Analysis &amp; results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Parkes et al. (2008)</td>
<td>Cross-sectional</td>
<td>818</td>
<td>Children</td>
<td>All</td>
<td>Representative, but cultural-specific</td>
<td>*Cerebral palsy by Gross Motor Function Classification System (GMFCS), visual acuity and Bimanual Fine Motor Function, hearing loss (decibels), seizures, pain (Child Health Questionnaire)</td>
<td>*Strengths and Difficulties Questionnaire: conduct, hyperactivity, emotion and peer problems</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Multivariate logistic regression: GMFCS score IV (severe limitations) OR 0.4 (95% CI 0.2–0.8), GMFCS score V (total assistance) OR 0.2 (95% CI 0.1–0.3) severe pain OR 2.7 (95% CI 1.5–4.6)</td>
<td><strong>2++</strong></td>
<td></td>
</tr>
<tr>
<td>Blacher &amp; McIntyre (2006)</td>
<td>Cross-sectional</td>
<td>282</td>
<td>Young adults</td>
<td>Moderate to profound</td>
<td>Representative, but cultural-specific</td>
<td>Scales of Independent Behavior – Revised (SIB-R), Problem Behavior Scale: internalising and externalising, and Reiss Screen for Maladaptive Behavior: aggression, SIB, sexual problems</td>
<td>ANOVA (cerebral palsy group compared to ID only, all less problems in CP): SIB-R general index P &lt; 0.05, externalised index P &lt; 0.05, internalised index not significantly different, asocial index not significantly different. Reiss: aggression P &lt; 0.05, SIB not significantly different, sexual problems not significantly different</td>
</tr>
</tbody>
</table>

**Table 4** Characteristics and level of evidence of studies on motor disorders

- **Exposure measure**: see Table 1
- **Outcome measure**: see Table 2
- **Level of evidence**: see Table 2

© 2011 The Authors. Journal of Intellectual Disability Research © 2011 Blackwell Publishing Ltd
people with ID, but a possible causal relationship was not investigated.

The severe functional limitations associated with motor disorders seem to prevent challenging behaviour, while the pain caused by the conditions is associated with increased challenging behaviour (Blacher & McIntyre 2006; high level of evidence).

Sensory impairments

We found three cross-sectional studies of sensory impairments: a well-conducted study, one with a high risk of bias and a non-analytical study (Table 5).

The well-conducted study from Sjoukes et al. (2009) found that visual impairment is not significantly related to challenging behaviour in adults who are visually impaired or blind compared to people without visual impairment. The second, lower-quality study indicates that persons with SIB were more often diagnosed with vision and hearing impairments (Wieseler et al. 1995). A likely bias is that more diagnostic examinations may have been performed in people with this behaviour. A high prevalence of visual impairment was noted in eye-poking children. Many of the eye-poking children also exhibited other types of SIB (Jan et al. 1994).

It has therefore not been proven that hearing impairment leads to more challenging behaviour in people with ID (low level of evidence). Visual impairment is not significantly associated with challenging behaviour (moderate level of evidence), but specific behaviours (such as SIB/eye-poking) may be related to visual impairment (low level of evidence).

Epilepsy

Our search traced eight studies of epilepsy. There were four well-conducted analytical studies and four analytical studies with a high risk of bias (Table 6).

There is no increased prevalence of physical aggression and other behavioural problems in adults with epilepsy compared to people without epilepsy (Matson et al. 1999; Espie et al. 2003; Tyrer et al. 2006; Matthews et al. 2008). Specific subgroups (people with additional visual impairment, motor handicaps, more severe and more frequent seizures and medication side effects, people with generalised EEG activity) may be more at risk for behavioural problems (Deb & Hunter 1991; Espie et al. 2003).

In a scientifically unsatisfactory study, Mendez et al. (1993) found that interictal violence was correlated with ID and psychopathology, but not with seizure variables.

Two low-quality studies looked at children. Children with epilepsy and ID had more behaviour problems than children with higher IQs, but this relationship was not significant after adjusting for seizure severity (Buelow et al. 2003). Lewis et al. (2000) found that children with epilepsy did not have more problem behaviour than those without epilepsy.

The group of epileptic patients as a whole does not therefore seem to exhibit more challenging behaviour, with the exception of some specific subgroups (moderate level of evidence).

Gastrointestinal disease

We identified eight studies of gastrointestinal disease. Seven were analytical studies with a high risk of bias or confounding, and one was a non-analytical study. Only one study had a prospective character, but it was uncontrolled. Six looked at gastro-oesophageal reflux disease (GORD) and two at infections with Helicobacter pylori (Table 7).

The study of Gössler et al. (2007) had a high risk of bias, but the results are nevertheless clinically relevant. Children with more gastro-oesophageal reflux showed significantly more agitation or SIB (as reported by parents). This was also true when there was recurrent reflux after treatment. Children with behavioural abnormalities had significantly higher levels of oesophageal inflammation.

Böhmer et al. (1999) found that rumination was more common in institutional residents with GORD. Behaviour problems (rumination, SIB, aggression, fear, screaming, restlessness) do not predict, but may co-occur with, GORD (Böhmer et al. 1997b). A significant association was found between reflux oesophagitis and problem behaviours or changed behaviour (rumination, aggression, fear, screaming, restlessness; Böhmer et al. 1997c). Rogers et al. (1992) found indications that rumination and meal-time challenging behaviours may be caused by dysphagia, GORD or aspiration. Adults
<table>
<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>n</th>
<th>Age</th>
<th>Level ID</th>
<th>Study population</th>
<th>Exposure measure</th>
<th>Outcome measure</th>
<th>Analysis &amp; results</th>
<th>Level of evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sjoukes et al. (2009)</td>
<td>Cross-sectional</td>
<td>269</td>
<td>Adults</td>
<td>Moderate to profound</td>
<td>Representative</td>
<td>***Visual functioning according to IASSID guidelines. Assessment of visual acuity, visual fields, contrast sensitivity, auto-refraction, skiascopy, strabismus</td>
<td>***Development Behaviour Checklist: disruptive/antisocial, self-absorbed, communication disturbance, anxiety, social relating</td>
<td>Multivariate linear regression: not significant</td>
<td>2+</td>
</tr>
<tr>
<td>Wieseler et al. (1995)</td>
<td>Cross-sectional</td>
<td>209</td>
<td>Children &amp; adults</td>
<td>Residence</td>
<td>Medical files (retrospectively)</td>
<td>SIB</td>
<td>Wilcoxon matched-pairs signed-ranks test: P &lt; 0.001</td>
<td>2-</td>
<td></td>
</tr>
<tr>
<td>Jan et al. (1994)</td>
<td>Case series</td>
<td>21</td>
<td>Children</td>
<td>Profound</td>
<td>Referred (visually impaired program)</td>
<td>Eye-poking</td>
<td>Frequent visual impairment in eye-poking and other SIB</td>
<td>3</td>
<td></td>
</tr>
</tbody>
</table>

Exposure and Outcome measure *, **, ***: see Table 1.
Level of evidence 2+, 2-, 3: see Table 2.
All = mild to profound ID.
Representative = study population is representative for ID population.
Referred = referred patients to a specialised centre/team.
Residence = study population is derived from a residence for people with ID.

ID, intellectual disabilities; IASSID, International Association for the Scientific Study of Intellectual Disabilities; SIB, self-injurious behaviour.
† No statistical analysis.
### Table 6 Characteristics and level of evidence of studies on epilepsy

<table>
<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>n</th>
<th>Age</th>
<th>Level ID</th>
<th>Study population</th>
<th>Exposure measure</th>
<th>Outcome measure</th>
<th>Analysis &amp; results</th>
<th>Level of evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Matthews et al. (2008)</td>
<td>Cross-sectional</td>
<td>318</td>
<td>Adults</td>
<td>Not described</td>
<td>Representative</td>
<td><strong>Epilepsy interview by trained nurse</strong></td>
<td><strong>Aberrant Behavior Checklist</strong></td>
<td>Comparison between epileptic and non-epileptic group (not different on level of functioning and psychiatric diagnoses). Mann–Whitney $U = 1254.0$, $P = 0.122$</td>
<td>2+</td>
</tr>
<tr>
<td>Tyrer et al. (2006)</td>
<td>Cross-sectional</td>
<td>3065</td>
<td>Adults</td>
<td>All</td>
<td>Representative</td>
<td><strong>Epilepsy by report of carer</strong></td>
<td><strong>Disability Assessment Schedule: physical aggression towards others</strong></td>
<td>Multivariate logistic regression epilepsy present: OR 0.95 (95% CI 0.74–1.21), $P = 0.67$</td>
<td>2+</td>
</tr>
<tr>
<td>Buelow et al. (2003)</td>
<td>Cross-sectional</td>
<td>164</td>
<td>Children</td>
<td>Borderline to mild (compared to normal and high IQ)</td>
<td>Representative</td>
<td><strong>Diagnosis of epilepsy, seizure severity scale for adults</strong></td>
<td><strong>Child Behaviour Checklist: internalising, externalising problems</strong></td>
<td>No comparison between presence and absence of epilepsy. Comparison between groups of different levels of ID: univariate analysis, total problem score higher in group with low IQ: $P = 0.0055$. After adjusting for seizure severity: $P = 0.094$</td>
<td>2–</td>
</tr>
<tr>
<td>Espie et al. (2003)</td>
<td>Cross-sectional</td>
<td>186</td>
<td>Adults</td>
<td>All</td>
<td>Referred (hospital-based epilepsy clinics, community ID teams, specialist teams)</td>
<td>Representative</td>
<td><strong>Neurologist diagnosis, seizure diaries and Epilepsy and Learning Disabilities Quality of Life Scale</strong></td>
<td>No comparison between presence and absence of epilepsy. Stepwise linear regression analysis: For irritability predictors: ambulant $P = 0.029$. For stereotypic behaviours: visual impairment $P = 0.038$.</td>
<td>2+</td>
</tr>
<tr>
<td>Lewis et al. (2000)</td>
<td>Cross-sectional</td>
<td>392</td>
<td>Children</td>
<td>All</td>
<td>Representative</td>
<td><strong>History of seizures</strong></td>
<td><strong>Developmental behaviour Checklist</strong></td>
<td>Multivariate analysis (MANCOVA): total behaviour problem score in epileptic group compared to non-epileptic group $P &gt; 0.05$</td>
<td>2–</td>
</tr>
<tr>
<td>Matson et al. (1999)</td>
<td>Cross-sectional</td>
<td>706</td>
<td>Adults</td>
<td>All</td>
<td>Residence</td>
<td><strong>Diagnosis of epilepsy by neurologist</strong></td>
<td><strong>Aberrant Behavior Checklist</strong></td>
<td>Univariate analysis (ANOVA): more aberrant behaviour in non-epileptic Group $F(1,705) = 19.1, P &lt; 0.001$</td>
<td>2–</td>
</tr>
</tbody>
</table>
Table 6 Continued

<table>
<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>n</th>
<th>Age</th>
<th>Level ID</th>
<th>Study population</th>
<th>Exposure measure</th>
<th>Outcome measure</th>
<th>Analysis &amp; results</th>
<th>Level of evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mendez et al. (1993)</td>
<td>Case–control</td>
<td>44–88</td>
<td>Adults</td>
<td>Mild to moderate</td>
<td>Referred (university-affiliated neurology clinic)</td>
<td>&quot;File study, measure not described&quot;</td>
<td>***Overt Aggression Scale: violence/aggression (verbal/physical/destructive/SIB)</td>
<td>No comparison between presence and absence of epilepsy. Comparison between violent group with epilepsy and non-violent group with epilepsy. Univariate analysis: more ID in the violent group. McNemar ( \chi^2 ); ( P &lt; 0.01 )</td>
<td>2−</td>
</tr>
<tr>
<td>Deb &amp; Hunter (1991)</td>
<td>Case–control</td>
<td>150–150</td>
<td>Adults</td>
<td>Mild to severe</td>
<td>Representative</td>
<td>***Epilepsy clearly defined, classification according to the International Classification of Epileptic Seizures</td>
<td>***Profile of Abilities and Adjustment (PAA) schedule (maladaptive behaviour section)</td>
<td>Groups comparable. Univariate analysis between epileptic (EP) and non-epileptic (NEP) groups: total PAA score and severe problem score not significantly different. EP less cooperative, ( Z = -2.21 ), ( P = 0.027 ). EP more echolalia, ( Z = -2.36 ), ( P = 0.018 ). Subgroup analysis: severe ID EP less aggressive than NEP (( Z = -1.97 ), ( P = 0.049 )). Single-type seizure EP less aggressive than NEP (( Z = -2.29 ), ( P = 0.022 )). Only slow background EEG activity EP less aggressive than NEP (( Z = -2.53 ), ( P = 0.011 )) and EP less overactive than NEP (( Z = -2.01 ), ( P = 0.044 )). Only generalised epileptiform EEG activity EP more temper tantrum than NEP (( Z = -2.47 ), ( P = 0.013 )) and EP more irritable than NEP (( Z = -2.42 ), ( P = 0.016 ))</td>
<td>2+</td>
</tr>
</tbody>
</table>

Exposure and Outcome measure * *, ** *, *** : see Table 1.
Level of evidence 2+, 2−: see Table 2.
All = mild to profound ID.
Representative = study population is representative for ID population.
Referred = referred patients to a specialised centre/team.
Residence = study population is derived from a residence for people with ID.
ID, intellectual disabilities; SIB, self-injurious behaviour.
<table>
<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>n</th>
<th>Age</th>
<th>Level ID</th>
<th>Study population</th>
<th>Exposure measure</th>
<th>Outcome measure</th>
<th>Analysis &amp; results</th>
<th>Level of evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gössler et al. (2007)</td>
<td>Uncontrolled prospective study</td>
<td>19</td>
<td>Children</td>
<td>Not described</td>
<td>Referred (paediatric department of university hospital)</td>
<td>&quot;Yes: 24-h pH monitoring and endoscopy&quot;</td>
<td>&quot;Auto-agression, agitation reported by caregivers&quot;</td>
<td>Univariate analysis. Children with behaviour abnormalities more GOR than without P &lt; 0.0004. SIB more GOR than agitation P &lt; 0.01. Children with behaviour abnormalities higher degree of inflammation than without P &lt; 0.05. Degree of inflammation not different between SIB and agitation P &gt; 0.05.</td>
<td>2-</td>
</tr>
<tr>
<td>Swender et al. (2006)</td>
<td>Case-control</td>
<td>60</td>
<td>Adults</td>
<td>Severe-profound</td>
<td>Residence</td>
<td>&quot;Medical records GORD (by pH testing or endoscopy)&quot;</td>
<td>&quot;Hand mouthing&quot;</td>
<td>Higher of frequency of GORD diagnosis in people with than without hand mouthing. $\chi^2 = 8.30, P &lt; 0.01$</td>
<td>2-</td>
</tr>
<tr>
<td>Wallace et al. (2002)</td>
<td>Cross-sectional</td>
<td>168</td>
<td>Adults</td>
<td>Not described</td>
<td>Representative</td>
<td>&quot;HP in blood &amp; faecal samples&quot;</td>
<td>&quot;Adaptive Behaviour Scale&quot;</td>
<td>Univariate analysis: more maladaptive behaviour in HP-positive than HP-negative group: stereotyped/ hyperactive P = 0.04, SIB P = 0.05. No difference: sexual behaviour P = 0.06, disturbing interpersonal behaviour P = 0.37</td>
<td>2-</td>
</tr>
<tr>
<td>Bohmer et al. (1999)</td>
<td>Cross-sectional</td>
<td>435</td>
<td>Children &amp; adults</td>
<td>Moderate to profound</td>
<td>Residence</td>
<td>&quot;24-h pH-metry and endoscopy&quot;</td>
<td>&quot;Physician report: rumination, SIB, aggression, fear, screaming, restlessness&quot;</td>
<td>Multivariate stepwise logistic regression GORD: rumination P = 0.001, other behaviours not significant</td>
<td>2-</td>
</tr>
<tr>
<td>Study</td>
<td>Design</td>
<td>n</td>
<td>Age</td>
<td>Level ID</td>
<td>Study population</td>
<td>Exposure measure</td>
<td>Outcome measure</td>
<td>Analysis &amp; results</td>
<td>Level of evidence</td>
</tr>
<tr>
<td>------------------</td>
<td>---------------</td>
<td>------</td>
<td>-------------</td>
<td>----------</td>
<td>------------------</td>
<td>------------------</td>
<td>----------------</td>
<td>------------------------------------------------------------------------------------</td>
<td>------------------</td>
</tr>
<tr>
<td>Böhmer et al.</td>
<td>Cross-sectional</td>
<td>338</td>
<td>Children &amp; adults</td>
<td>Moderate to profound</td>
<td>Residence</td>
<td>***Serum test HP</td>
<td>**Staff report: rumination</td>
<td>Univariate analysis: rumination in HP-positive patients, compared to HP-negative patients: OR 2.0 (95% CI 1.07–3.64) ( P = 0.04 )</td>
<td>2–</td>
</tr>
<tr>
<td>(1997a)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Böhmer et al.</td>
<td>Cross-sectional</td>
<td>110</td>
<td>Children &amp; adults</td>
<td>Moderate to profound</td>
<td>Residence</td>
<td>***24-h pH-metry and endoscopy</td>
<td>**Staff report: rumination, SIB, aggression, fear, screaming, restlessness</td>
<td>Univariate analysis: comparison of frequency of behaviour problems in patients with GORD and without: all not significant</td>
<td>2–</td>
</tr>
<tr>
<td>(1997b)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Böhmer et al.</td>
<td>Cross-sectional</td>
<td>1687</td>
<td>Children &amp; adults</td>
<td>Moderate to profound</td>
<td>Residence</td>
<td>***24-h pH-metry and endoscopy (retrospective file study)</td>
<td>**Physician report: rumination, SIB, aggression, fear, screaming, restlessness</td>
<td>Univariate analysis: comparison of frequency of behaviour problems in patients with reflux oesophagitis and without: rumination ( P &lt; 0.0001 ), changed behaviour ( P = 0.01 )</td>
<td>2–</td>
</tr>
<tr>
<td>(1997c)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rogers et al.</td>
<td>Case series</td>
<td>23</td>
<td>Adults</td>
<td>Profound</td>
<td>Residence</td>
<td>***Modified barium swallows and esophagrams</td>
<td>**Staff supervisors: regurgitation Psychologist reports: other behaviour problems</td>
<td>High prevalence of dysphagia and gastro-oesophageal abnormalities in rumination†</td>
<td>3</td>
</tr>
<tr>
<td>(1992)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Exposure and Outcome measure *, **, ***: see Table 1.
Level of evidence 2–, 3: see Table 2.
All = mild to profound ID.
Representative = study population is representative for ID population.
Referred = referred patients to a specialised centre/team.
Residence = study population is derived from a residence for people with ID.
ID, intellectual disabilities; SIB, self-injurious behaviour; GORD, gastro-oesophageal reflux disease; HP, Helicobacter pylori.
† No statistical analysis.
who engaged in hand mouthing were diagnosed with GORD more often (Swender et al. 2006).

Current H. pylori infection was associated with more problem behaviour (stereotyped, hyperactive and self-abusive). H. pylori infection was also associated with a lower level of ID. This was not taken into account in the analysis of the correlation with problem behaviour (Böhmer et al. 1997a; Wallace et al. 2002).

Gastro-oesophageal reflux disease may therefore contribute to behaviour difficulties such as rumination, SIB and agitation, but this is insufficiently substantiated (low level of evidence).

Sleep disorders

Seven studies of sleep disorders were found: one well-conducted, five analytical with a high risk of bias or confounding, and one non-analytical study (Table 8).

In the well-conducted study, children with sleep problems had more daytime problem behaviours (irritability, stereotypy, hyperactivity, aggression, screaming, temper tantrum, non-compliance and impulsivity) than children without sleep problems. When factors for specific sleep problems were analysed, after correction for age and level of ID, only irritability was associated with severe sleep problems. The cross-sectional design implies that it is not possible to determine whether the problem behaviours or the sleep problems are the cause (Didden et al. 2002).

In studies with a high risk of bias, looking at both children and adults, the subjects with problem behaviours (SIB) had more sleep disturbance than those without problem behaviours (Piazza et al. 1996; Symons et al. 2000), and people with sleep problems displayed more, and more severe, problem behaviour (stereotypes, irritability, SIB, aggression, temper tantrums, screaming) than those without sleep problems. It is unclear whether sleep problems cause the behavioural problems or vice versa, or if both have a shared neurobiological base (Quine 1991; Chaney et al. 1994; Wiggs & Stores 1996; Brylewski & Wiggs 1999).

Sleep problems are therefore associated with daytime challenging behaviour, but the nature of the relationship remains unestablished (moderate level of evidence).

Dementia

We found two non-analytical studies of dementia (which did not relate specifically to Down syndrome only; Table 9).

In people with dementia, behaviour problems (agression, temper tantrums, pica, SIB, screaming, wandering, repetitive behaviours) are seen, and may arise early in the disease process (Duggan et al. 1996; Cooper 1997; very low level of evidence).

Others

We found three articles about conditions not mentioned previously (Table 10).

In a well-conducted study of pain in children with ID, Breau et al. (2003) showed that children with SIB did not express pain differently. Children with chronic pain did not have more SIB than children without chronic pain, but the locations were different (near the site of pain instead of mostly to the head and hand).

A non-analytical study of male sex offenders with ID found that many had a distinctive physical disability that might have contributed to the behaviour (Day 1994).

In a non-analytical study of the menstrual cycle in women with SIB, Taylor et al. found that SIB might be exacerbated during certain phases of the menstrual cycle (Taylor et al. 1993; very low level of evidence).

Discussion

This systematic review of physical conditions associated with challenging behaviour in children and adults with ID identified 11 well-conducted studies that found significant and independent associations with urinary incontinence, pain related to cerebral palsy and chronic sleep problems. Visual impairment is significantly associated with SIB. Because of the cross-sectional design of all the studies, no firm conclusions can be drawn about the causative character of the physical conditions discussed. Longitudinal studies are the only way to complete the evidence. No association was found with hearing impairment, bowel incontinence, mobility impairment or epilepsy. Twenty-one analytical studies of unsatisfactory quality and 13 non-analytical studies...
<table>
<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>n</th>
<th>Age</th>
<th>Level ID</th>
<th>Study population</th>
<th>Exposure measure</th>
<th>Outcome measure</th>
<th>Analysis &amp; results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Didden et al. (2002)</td>
<td>Cross-sectional</td>
<td>286</td>
<td>Children</td>
<td>All</td>
<td>Representative</td>
<td><strong>Simonds &amp; Parraga Sleep Questionnaire</strong></td>
<td><strong>Aberrant Behavior Checklist</strong></td>
<td>Mann-Whitney U, sleep problem compared to no sleep problem: irritability $Z = -3.93$ $P &lt; 0.001$, stereotypy $Z = -3.60$ $P &lt; 0.001$, hyperactivity $Z = -3.73$ $P &lt; 0.001$, SIB $Z = -1.11$ $P = 0.266$, aggression $Z = -3.15$ $P &lt; 0.01$, screaming $Z = -2.90$ $P &lt; 0.01$, temper tantrums $Z = -3.68$ $P &lt; 0.001$, non-compliance $Z = -4.31$ $P &lt; 0.001$, impulsivity $Z = -2.09$ $P &lt; 0.05$. Multivariate stepwise logistic regression analysis for specific sleep problems: only irritability significant correlation with severe sleep problems: $Wald \chi^2 = 10.23$ $P &lt; 0.01$.</td>
</tr>
<tr>
<td>Symons et al. (2000)</td>
<td>Case-control</td>
<td>60</td>
<td>Adults</td>
<td>Profound</td>
<td>Residence</td>
<td><strong>Sleep observations by staff</strong></td>
<td><strong>SIB (as reported by staff)</strong></td>
<td>SIB less sleep than people without SIB: Wilcoxon signed-rank test: $Z = 2.31$ $P &lt; 0.02$, and more variability in sleep pattern: $\chi^2 &lt; 0.01$.</td>
</tr>
<tr>
<td>Brylewski &amp; Wiggs (1999)</td>
<td>Cross-sectional</td>
<td>205</td>
<td>Adults</td>
<td>All</td>
<td>Representative</td>
<td><strong>Simonds &amp; Parraga sleep questionnaire</strong></td>
<td><strong>Aberrant Behavior Checklist</strong></td>
<td>t-test for differences between groups with and without sleep problems: irritability $t = -3.76$ $P &lt; 0.001$, stereotypies $t = -3.07$ $P &lt; 0.01$, hyperactivity $t = -2.62$ $P = 0.01$, aggression/temper tantrums $t = -3.12$ $P &lt; 0.01$, non-compliance $t = -1.81$ $P = 0.073$, SIB $t = -2.83$ $P &lt; 0.01$, screaming $t = -3.26$ $P = 0.001$.</td>
</tr>
<tr>
<td>Wiggs &amp; Stores (1996)</td>
<td>Cross-sectional</td>
<td>486</td>
<td>Children</td>
<td>Severe</td>
<td>Representative</td>
<td><strong>Simonds &amp; Parraga sleep questionnaire</strong></td>
<td><strong>Aberrant Behavior Checklist</strong></td>
<td>t-test for differences between groups with and without sleep problems: irritability $t = 4.43$ $P &lt; 0.001$, stereotypies $t = 3.69$ $P &lt; 0.001$, hyperactivity $t = 4.18$ $P &lt; 0.001$, SIB $t = 2.76$ $P &lt; 0.01$, aggression/temper tantrums $t = 5.22$ $P &lt; 0.001$, screaming $t = 5.54$ $P &lt; 0.001$, non-compliance $t = 3.60$ $P &lt; 0.001$, impulsivity 2.90 $P &lt; 0.01$.</td>
</tr>
<tr>
<td>Study</td>
<td>Design</td>
<td>n</td>
<td>Age</td>
<td>Level ID</td>
<td>Study population</td>
<td>Exposure measure</td>
<td>Outcome measure</td>
<td>Analysis &amp; results</td>
</tr>
<tr>
<td>-----------------------</td>
<td>-----------------</td>
<td>-----</td>
<td>-----------------</td>
<td>----------------</td>
<td>----------------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------------------</td>
<td>----------------------------------------------------------------------------------</td>
<td>---------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Piazza et al. (1996)</td>
<td>Cross-sectional</td>
<td>51</td>
<td>Children &amp; young adults</td>
<td>All</td>
<td>Referred (inpatient unit for people with severe behaviour problems)</td>
<td><strong>Sleep observations</strong></td>
<td>Behaviour reports by psychologist</td>
<td>Less sleep in children with ID and behaviour problems than peers of the same age†</td>
</tr>
<tr>
<td>Chaney et al. (1994)</td>
<td>Cross-sectional</td>
<td>40</td>
<td>Adults</td>
<td>Moderate to profound</td>
<td>Residence</td>
<td>*<strong>Sleep observations</strong></td>
<td>***Client Development Evaluation report: aggression, SIB, destructiveness, reaction to frustration, stereotypy, hyperactivity, temper tantrums, unacceptable social behaviour</td>
<td>Fisher’s exact probability test patients with sleep disturbance compared to those without more stereotypic behaviour: $P = 0.01$, SIB: $P = 0.056$</td>
</tr>
<tr>
<td>Quine (1991)</td>
<td>Cross-sectional</td>
<td>166</td>
<td>Children</td>
<td>Severe Representative</td>
<td>***Sleep index and settling and waking problems from the Behaviour Screening Questionnaire</td>
<td>***Behaviour Screening Questionnaire and Disability Assessment Schedule</td>
<td>$\chi^2$ between children with and without sleep problems: management problems $P &lt; 0.001$, activity $P &lt; 0.001$, concentration $P &lt; 0.001$, attention seeking $P &lt; 0.01$, sexual problems $P &lt; 0.001$, runs away $P &lt; 0.001$, interferes $P &lt; 0.001$, destructive $P &lt; 0.01$, pica $P &lt; 0.01$, swears $P &lt; 0.01$, disruptive $P &lt; 0.01$, temper tantrums $P &lt; 0.05$, problems with peers $P &lt; 0.05$, habits $P &lt; 0.001$, repetitive activities $P &lt; 0.05$, echolalia $P &lt; 0.05$</td>
<td>2–</td>
</tr>
</tbody>
</table>

Exposure and Outcome measure *, **, ***: see Table 1.
Level of evidence 2+, 2–, 3: see Table 2.
All = mild to profound ID.
Representative = study population is representative for ID population.
Referred = referred patients to a specialised centre/team.
Residence = study population is derived from a residence for people with ID.
ID, intellectual disabilities; SIB, self-injurious behaviour.
† No statistical analysis.
Table 9 Characteristics and level of evidence of studies on dementia

<table>
<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>n</th>
<th>Age (years)</th>
<th>Level ID</th>
<th>Study population</th>
<th>Exposure measure</th>
<th>Outcome measure</th>
<th>Analysis &amp; results</th>
<th>Level of evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duggan et al. (1996)</td>
<td>Case series</td>
<td>12</td>
<td>≥48</td>
<td>Mild to severe</td>
<td>Referred (psychiatrists for people with ID)</td>
<td>***If available dementia diagnosis by psychiatrist in medical files and interview with carers</td>
<td>**Past Behavioural History Inventory</td>
<td>Behavioural problems (pica, aggression, SIB, screaming, wandering, repetitive behaviours) can occur early in dementia†</td>
<td>3</td>
</tr>
</tbody>
</table>

Exposure and Outcome measure * *, ** *, *** : see Table 1.
Level of evidence 3: see Table 2.
All = mild to profound ID.
Representative = study population is representative for ID population.
Referred = patients identified by specialised consultants.
ID, intellectual disabilities; SIB, self-injurious behaviour.
† No statistical analysis.
<table>
<thead>
<tr>
<th>Study</th>
<th>Design</th>
<th>n</th>
<th>Age</th>
<th>Level ID</th>
<th>Study population</th>
<th>Exposure measure</th>
<th>Outcome measure</th>
<th>Analysis &amp; results</th>
<th>Level of evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Breau et al.</td>
<td>Cross-sectional</td>
<td>101</td>
<td>Children</td>
<td>All</td>
<td>Referred (tertiary paediatric centre)</td>
<td>***Pain: Non-communicating Children’s Checklist – Revised</td>
<td>**SIB: Behavior Problem Inventory</td>
<td>MANOVA: not significant for pain expression between children with and without SIB. t-test: children with chronic pain compared to those without; less body surface, ( P = 0.01 ), fewer body sites, ( P = 0.04 )</td>
<td>2+</td>
</tr>
<tr>
<td>Day (1994)</td>
<td>Case series</td>
<td>47</td>
<td>Adolescents &amp; adults</td>
<td>Borderline to moderate</td>
<td>Referred (hospital department of psychiatry)</td>
<td>**Medical files (retrospectively)</td>
<td>**Sexual offences/ incidents</td>
<td>51% had a distinctive physical disability (speech deficits, dysmorphias, sensory impairments, epilepsy)†</td>
<td>3</td>
</tr>
<tr>
<td>Taylor et al.</td>
<td>Case series</td>
<td>9</td>
<td>Adolescents &amp; adults</td>
<td>Severe to profound</td>
<td>Residence</td>
<td>**Menstrual cycle</td>
<td>**Client Development Evaluation Report: SIB</td>
<td>Exacerbation of SIB during early and late follicular phases of the menstrual cycle†</td>
<td>3</td>
</tr>
</tbody>
</table>

Exposure and Outcome measure *, **, ***: see Table 1.
Level of evidence 2+, 3: see Table 2.
All = mild to profound ID.
Referred = referred patients to a specialised hospital/tertiary centre.
Residence = study population is derived from a residence for people with ID.
ID, intellectual disabilities; SIB, self-injurious behaviour.
† No statistical analysis.
or case series, usually based on file data, suggest associations with GORD, dysphagia, dementia, menstrual cycle phases and specific sub-types of epilepsy. However, so far, these are insufficiently substantiated.

Reports listing undiagnosed or untreated conditions found during the diagnostic work-up of persons with challenging behaviours (Ryan & Sunada 1997; Kastner et al. 2001) have only limited value, as it is well known that even severe undiagnosed conditions are found in any group of persons with ID (van Schrojenstein Lantman-de Valk & Walsh 2008). This results from the ineffective communication of subjective symptoms by these persons and by a lack of training in medical issues among caring staff.

Four medical conditions have been proven to be associated with challenging behaviour. Urinary incontinence is significantly correlated with aggressive behaviour. As the authors in question point out, it remains unclear whether there is a shared underlying mechanism (e.g. autonomic sympathetic discharge), whether the incontinence contributes to aggression (because people are ashamed or experience discomfort) or whether the aggression contributes to the incontinence (Cooper et al. 2009b).

Second, visual impairment is significantly correlated with SIB only. The cross-sectional character of the studies means that the direction of this relationship has not been established. Visual impairment may contribute to the behaviour (people express communication difficulties with problem behaviour, or engage in eye-poking in order to get visual stimulation); alternatively, visual impairment may be caused by the tissue damage resulting from SIB.

Third, increased behavioural difficulties are seen only if cerebral palsy is complicated by severe pain (Parkes et al. 2008). Finally, the shared conclusion in studies of sleep disorders is that people with behavioural problems have more sleep disturbance (Quine 1991; Chaney et al. 1994; Piazza et al. 1996; Wiggs & Stores 1996; Brylewski & Wiggs 1999; Symons et al. 2000). It remains unclear whether the association is because of a common underlying neurochemical mechanism accounting for both factors, and whether sleep disturbance contributes to behaviour problems, or vice versa (Symons et al. 2000). These findings are supported by single-case experimental studies, in which problem behaviours are exacerbated in people who are naturally deprived of sleep (Kennedy & Meyer 1996; O’Reilly & Lancioni 2000).

A few conditions that appear clinically relevant have been insufficiently investigated. Almost all studies of the behavioural effects of gastrointestinal disease are side measurements of other studies. Dysphagia, GORD and H. pylori infections are frequent in rumination cases (Rogers et al. 1992; Böhmer et al. 1997a), which indicates that there should be diagnostic testing for these conditions in people with rumination. Changes in behaviour may be caused by oesophageal pain, colic and discomfort resulting from GORD (Gössler et al. 2007) and reflux oesophagitis (Böhmer et al. 1997c).

Dementia can result in behavioural changes, even at an early stage (Duggan et al. 1996; Cooper 1997). One study indicates that distinctive physical features may cause aberrant behaviour, possibly resulting from low self-esteem (Day 1994). The menstrual cycle may affect SIB, either through hormonal changes or pain and discomfort (Taylor et al. 1993). The role of pain requires further study, because there is no proof that chronic pain causes more SIB (Breau et al. 2003), even though there is a proven link between severe pain and behaviour in children with cerebral palsy (Parkes et al. 2008). Acute and chronic pain may very well be expressed by challenging behaviour (Dubois et al. 2010).

In certain physical conditions, it has not been possible to prove a causal relationship. Motor disorders are not correlated with more problem behaviours (Blacher & McIntyre 2006; Jones et al. 2008; Parkes et al. 2008). This may be because people with impaired mobility may receive higher levels of support from carers (to meet their disability needs). Furthermore, the physical disabilities may preclude problem behaviour in these people (Jones et al. 2008). Nor has hearing impairment been proven to cause more problem behaviours (Wieseler et al. 1995; Jones et al. 2008; Cooper et al. 2009a,b). However, many studies used file data and it is known that hearing impairment frequently goes unnoticed (Evenhuis et al. 2001; Meuwese-Jongejeugd et al. 2008), so this association should be investigated further.

Conclusions in studies of epilepsy are unambiguous. Generally, the population with epilepsy does not have more behaviour problems than those
without epilepsy (Deb & Hunter 1991; Matson et al. 1999; Lewis et al. 2000; Buelow et al. 2003; Espie et al. 2003; Tyrer et al. 2006; Jones et al. 2008; Matthews et al. 2008; Cooper et al. 2009a,b). However, it has been suggested that specific subgroups (e.g. people with generalised epileptic activity, more severe or frequent seizures, medication side effects and co-morbidity) may be prone to more behavioural difficulties (Deb & Hunter 1991; Espie et al. 2003).

A possible explanation of why associations are difficult to detect is that, in larger studies, the groups of participants were very heterogeneous. This may have reduced the effect sizes.

There are many medical conditions (e.g. infectious disease, migraine, menopause, allergy, and cardiac and pulmonary disease) where it is conceivable that pain or discomfort could lead to challenging behaviour. Particularly, people with more severe forms of ID, with less capacity to communicate their discomfort, may express this through altered behaviour. However, this review failed to find studies of many of these conditions. This does not mean that there is no correlation between these conditions and challenging behaviour; it means that further investigation is required. Areas warranting urgent attention in future research are gastrointestinal disease (GORD, but also constipation), infectious disease (e.g. ear infections, which can cause pain), hormonal influences (menstrual cycle and thyroid function), dental disease and cardiopulmonary disease. The fact that the life expectancy of the population with ID is rapidly increasing means that research is needed into the behavioural effects of physical deterioration and menopause in older age.

Only a few studies directly address the question of which physical conditions may cause problem behaviour and, with a few exceptions, these were generally low-quality studies. More often, behaviour was measured in conjunction with a single physical condition, or behaviour was described, but as part of a larger study of the condition. Moreover, the design of almost all the studies was cross-sectional. The implication is that the nature of the associations found in these studies cannot be established. This makes firm conclusions about causal relationships difficult but some suggestions for clinical practice can be made.

When people with ID and challenging behaviours are examined, urinary incontinence, visual impairment, sleep problems and pain should be considered. In people who engage in rumination, hand mouthing or mealtime challenging behaviour, diagnostic testing should be performed looking at dysphagia, GORD and H. pylori infections. Other conditions that might be considered are early dementia, discomfort in certain menstrual cycle phases and specific sub-types of epilepsy (people with more severe and more frequent seizures, medication side effects and generalised EEG activity).

The present review highlights the role that medical conditions can play in challenging behaviour and the need for evaluating those conditions in clinical practice. It also reveals major gaps in the evidence, because many studies are of low quality and many physical illnesses have not been investigated.

There is a strong need for comprehensive high-quality longitudinal research, with clearly defined measures of both physical conditions and behavioural disorders, to establish firm evidence as a basis for clinical guidelines for the prevention, differential diagnosis and treatment of challenging behaviour.

Acknowledgements

This study was performed at the request of the Centre for Consultation and Expertise, the Netherlands, as part of the expertise project ‘Medical conditions and challenging behaviour’.

References


*Accepted 18 January 2011*