Screening of dementia indicating signs in adults with intellectual disabilities

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Abstract

Background: In intellectual disability, the cognitive delay is observed during developmental age, whereas in dementia, cognitive decline occurs during post-developmental period. So far, the risk of dementia in people with intellectual disability, excluding those with Down syndrome, is poorly known.

Method: We screened dementia signs in a study group of 230 adults (34–80 years of age) with the help of the British Present Psychiatric State—Learning Disabilities assessment.

Results: Of the study members, 42% showed two or more signs. The overall frequency of symptoms did not differ between age groups. The number of individuals with a genetic syndrome or disease manifesting with a shortened lifespan was greater in the younger age groups when compared to the older age groups.

Conclusion: People with an intellectual disability represent numerous rare syndromes with comorbidities. It seems that dementia signs may affect any age groups of adults with intellectual disability.

KEYWORDS
ageing, dementia, intellectual disability, memory disorders, Present Psychiatric State—Learning Disabilities assessment (PPS-LD)

INTRODUCTION

People with intellectual disability are the largest single disability group. They require other people’s assistance as well as public services over their entire lifespan, and therefore, the impact of intellectual disability on society is remarkable. Intellectual disability research has focused mainly on young people. Over the last several decades, the average life expectancy of people with intellectual disability has increased considerably and a greater number of senior-aged people with intellectual disability live in our communities (Arvio et al., 2016). Consequently, dementia has become a contributing factor for health also in this population (Sheehan et al., 2014, 2015). It is well known that Down syndrome is associated with an increased risk of early-onset Alzheimer’s disease (Lott & Head, 2019). It has been suggested that an increased risk of dementia applies also to persons with intellectual disability without Down syndrome aged 60+ compared to the general population (Silverman et al., 2013, Strydom et al., 2013, Takenoshita et al., 2020).

Dementia needs to be distinguished from intellectual disability even though both intellectual disability and dementia result from dysfunctions of the cerebral cortex, and the cause of both disorders can be genetic, acquired or multifactorial. Intellectual disability is diagnosed during developmental years (under the age of 18) when the intellectual and adaptive skills of an individual are two standard deviations below what is expected for the age-matched group. Dementia refers to a clinical syndrome at adulthood, which is...
characterized by a progressive cognitive decline that interferes with the person’s ability to function independently. Symptoms of dementia are gradual, persistent and progressive. Intellectual disability as well as dementia affects many domains; the social, communication, cognitive, adaptive and often motor skills of an affected person are weaker than his/her non-disabled and non-demented peers.

Intellectual disability is caused by numerous aetiologies and can also be associated with several risk factors for dementia. The most common genetic intellectual disability syndrome is Down syndrome, the most common acquired syndrome is cerebral palsy–intellectual disability syndrome (CP-IDS), and the most common multifactorial syndrome is intellectual disability comorbid with autism spectrum disorder (ASD-IDS) (Arvio & Aaltonen, 2011). Dementia risk factors affecting persons with intellectual disability especially can include, for example, brain dysfunction caused by brain damage, structural brain anomaly or persistent epileptic seizures. Some intellectual disability syndromes are associated with overweight, while other persons with intellectual disability may suffer from malnutrition due to swallowing problems or spasticity. Persons with intellectual disabilities may also have limited access to exercise and other cultural activities, as they are dependent on others’ assistance in their daily life. All these factors may contribute to the development of dementia.

Since the early 1990s, it has been recognized that adults with intellectual disability who are affected by dementia need special care (Janicki & Keller, 2015; McCarthy & Mullan, 1996). Our earlier studies suggest that the dementia rates in intellectual disability excluding Down syndrome and compared to the general population may be either higher, lower or similar depending on the aetiology of intellectual disability (Arvio, 2016; Arvio & Luostarinen, 2016; Sauna-aho et al., 2018, 2019, 2020). The purpose of the present cross-sectional study was to screen dementia signs in different age groups of adults with intellectual disability.

2 | SUBJECTS AND METHOD

The study population included subjects who fulfilled the predetermined criteria. They were resident in a geographically defined area, the catchment area of four joint authorities in northern Finland, were over 34 years of age and had an intellectual disability. The age of 34 was chosen since in Down syndrome, the signs indicating dementia are known to appear soon after the age of 35 (Arvio & Bjelogrlic-Laakso, 2017).

First, the study design was evaluated and approved by the institutional review board of the Oulu University Hospital. Three research nurses then informed municipal disability service officers of the four joint authorities of the study. These officers mailed written information of the study and a consent form with a return envelope to eligible subjects living in their catchment area. Subjects who themselves or whose proxy had given written consent were face-to-face interviewed along with their close care providers at home by the three research nurses using the Finnish translation of the British Present Psychiatric State—Learning Disabilities scale (PPS-LD, 27 items) (Cooper, 1997a, 1997b; Mölsä, 2001). PPS-LD has been widely used in several of our earlier studies for 20 years (Arvio et al., 2013;
Arvio & Bjelogrlic-Laakso, 2017; Sauna-aho et al., 2018). Statistical significance of linearity across age groups of the dementia sign score was evaluated by using the bootstrap type analysis of variance with an appropriate contrast (orthogonal). Correlations were computed with Spearman’s correlation coefficients with bootstrapped 95% confidence intervals.

3 | RESULTS

Of the eligible 467 subjects, 230 (49%, 128 males and 102 females) were willing to participate.

In total, 70 (30%) had a specific aetiological diagnosis. There were 32 individuals with Down syndrome, 19 with CP-IDS and 19 with other genetic syndromes. Other genetic disorders included three people with aspartylglucosaminuria, seven with fragile X, two with Catch-22, two with Klinefelter and one syndrome from each of Cornelia de Lange, Prader–Willi, Rubinstein–Taybi, Smith–Lemli–Opitz, Rett and Williams. Eighteen (8%) had been diagnosed with ASD-IDS. The age distribution of the study group is presented in Figure 1. The average age of the study group was 51 years; 53 (23%) individuals had reached the age of 60 and one the age of 80. Eight people with Down syndrome had been diagnosed with Alzheimer’s disease prior to the study.

The majority (85%) were on continuous medications; one-third used antiepileptic and one-third psychiatric drugs. All but one individual with ASD-IDS were on psychiatric drug treatment.

Table 1 shows the distribution of the 27 dementia symptoms. The most common symptoms were loss of energy, reduced self-care skills, forgetfulness and diurnal mood variation. Individuals with a suspected memory disorder were referred for further neuropsychological and medical examinations.

Of the 230 subjects, 104 (45%) did not manifest any of the 27 dementia symptoms. The average age of this zero-score group was 48 years. Further, the average age of 92 individuals with 1–4 symptoms was 54 years, of 25 individuals with 5–9 symptoms 51 years and of 10 with 9+ symptoms 50 years.

We divided the study group into five age groups. In the youngest age group (34–39 years), 41% had a verified genetic aetiological diagnosis, while the respective percentage in the oldest age group (70+ years) was 5% (Figure 1, footnotes). The division of dementia sign scores of the five age groups is presented in Figure 1. The scores were not significantly higher in older age groups when compared to younger age groups.

Further, the dementia sign scores were compared between three subgroups: between those with Down syndrome (N = 32), CP-IDS (N = 19) and ASD-IDS (N = 18) (Figure 2). The subgroups of Down syndrome and ASD-IDS manifested significantly higher scores than the CP-IDS subgroup (Figure 2). However, the scores of the ASD-IDS subgroup did not correlate with age as did the scores of Down syndrome subjects. The scores of the eight persons with Down syndrome and Alzheimer disease diagnosis ranged from 2 to 12 (mean 6).

4 | DISCUSSION

Diagnosing dementia in a patient with intellectual disability is very important, because even a small decline in functional ability may be detrimental to a person’s daily living. It may mean, for example, a premature need to move into a nursing home providing 24/7 assistance if the appropriate interventions (depending on the cause of dementia) are not initiated on time. Relatives, other carers, psychologists and physicians familiar with memory disorders play a critical role in early recognition of dementia.

The majority of adults with intellectual disability live in nursing homes, where regular follow-up of functional ability forms a central tool in detecting the first signs of dementia. When the first symptoms are noted, somatic disorders such as B12 vitamin deficiency, cystitis and thyroid dysfunction need to be excluded (or treated if found) by laboratory tests. During evaluations, it is important to

### Table 1 Signs indicating dementia in the study group

| Sign                               | Study group (N = 230) |
|------------------------------------|-----------------------|
| Autonomic anxiety                   | 17 (7.5%)             |
| Change in appetite                  | 7 (3%)                |
| Changed sleep pattern               | 20 (9%)               |
| Coarsening of personality           | 20 (9%)               |
| Confusion                           | 8 (3.5%)              |
| Delusions                           | 6 (2.5%)              |
| Diurnal mood variation              | 24 (10.5%)            |
| Forgetfulness                       | 24 (10.5%)            |
| Forgetting people's names           | 22 (9.5%)             |
| Geographical disorientation         | 9 (4%)                |
| Impaired understanding              | 13 (5.5%)             |
| Irritability                        | 23 (0%)               |
| Loss of concentration               | 17 (7.5%)             |
| Loss of energy                      | 36 (16%)              |
| Loss of literacy skills             | 7 (3%)                |
| Misery                              | 13 (5.5%)             |
| Onset of or increase                |                       |
| In other maladaptive behaviour      | 16 (7%)               |
| In physical aggression              | 17 (7.5%)             |
| In verbal aggression                | 9 (4%)                |
| Of fearfulness                      | 15 (6.5%)             |
| Reduced quantity of speech          | 18 (8%)               |
| Reduced self-care skills            | 40 (17.5%)            |
| Social withdrawal/reduced social interaction | 10 (4.5%) |
| Tearfulness                         | 17 (7.5)              |
| Temporal disorientation             | 8 (3.5%)              |
| Weight change                       | 21 (9.5%)             |
| Worry                               | 16 (7%)               |
recognize possible depression, sleep apnoea and other sleep disorders all of which may have a negative impact on memory. Different forms of epilepsy may start at any age and influence vitality. The possible negative role of medication in functionality and memory decline should not be overlooked neither.

This cross-sectional study evaluated signs indicating dementia in different age groups of persons with intellectual disability. When the dementia sign scores of the whole study group were analysed, somewhat surprisingly, the prevalence of dementia symptoms in the over-70-year-old age group was not significantly greater as compared to the under-40-year-old age group. Eight (3%) study members already had a verified memory disorder diagnosis, and 126 (55%) showed dementia sign/signs warranting further medical studies. A common dilemma in cross-sectional studies may explain the unexpected outcome of our study: the different age groups were not homogenous in terms of aetiology. The younger age group comprised individuals with severe intellectual disability syndromes, while the over-70-year-old age group included one man with fragile X and two with Klinefelter syndrome, which is not classified as an intellectual disability syndrome but which in these particular cases were associated incidentally with intellectual disability. Thus, it may be justified to claim that one cannot make any reliable conclusions on whether intellectual disability (without known specific aetiology) increases the risk of dementia or not.

Our earlier cross-sectional as well as longitudinal studies have focused on three genetic syndromes, that is Down, fragile X and Williams syndromes (Arvio, 2016; Arvio & Luostarinen, 2016; Sauna-aho et al., 2018, 2019, 2020). The association between Down syndrome and Alzheimer’s disease has been known for a long time (Takashima, 1997). However, Down syndrome is a risk factor also for vascular dementia. Moyamoya disease, a rare cerebrovascular disorder, is prevalent in people with Down syndrome, exposing them to an increased risk of cerebral haemorrhage and vascular dementia (Vila-Herrero et al., 2004). Many adults with Williams syndrome suffer from metabolic syndrome exposing them to vascular dementia (Sauna-aho et al., 2019). Fragile X syndrome does not appear to increase the risk of any type of memory disorder (Arvio, 2016; Sauna-aho et al., 2018, 2020).

The ASD-IDS group was found to have high dementia sign scores but without an age correlation as seen in the Down syndrome group. There are several clinical and aetiological factors which may explain this somewhat unexpected finding. Firstly, diagnostics is vague taken that autism spectrum disorders are caused by multiple genetic and environmental factors. As the research subjects have not been exome sequenced, some subjects may have some as yet undetected syndrome associated with early dementia. Secondly, the differential diagnostics of self-destructive and aggressive behaviours often related to ASD-IDS is challenging. Behavioural symptoms may be due to unrecognized psychosocial/environmental factors or some psychotic disorder. Frequently, these patients have been on multiple antipsychotic and other neuropsychiatric medication since their childhood or early adolescence (Bjelogrlic-Laakso et al., 2020). The role of long-term pharmacotherapy in persistent behavioural symptoms, and its effects on developing brains are largely unknown. One explanation for dementia signs observed already at an unusually early age in patients with autism spectrum disorder can in certain cases be the repeated head banging resulting in boxer’s dementia (Hof et al., 1991).

In the clinical context, the intellectual disability population is an extremely heterogeneous group in terms of the cause, level of severity and associated comorbidities of the disability, which sets a considerable challenge for research into intellectual disabilities and dementia. From an aetiological perspective, our current study group may reflect
a typical intellectual disability population that a physician encounters in everyday practice. Along with the most common genetically caused syndrome, we focused on the most common acquired and common multifactorial intellectual disability syndromes representing a typical intellectual disability population. The lack of age-related increase in dementia signs observed in this population confirms our earlier findings on the importance of a specific aetiological diagnosis (Arvio, 2016; Arvio & Luostarinen, 2016; Sauna-aho et al., 2018, 2019, 2020).

The strength of our study is that an experienced nurse interviewed the study members and their close care providers and when deemed necessary referred them for further examinations. The main limitation is that we do not know how many study members actually suffered from a memory disease.

Conclusion: Our cross-sectional study showed that dementia indicating symptoms are almost as common in under-40-year-olds as in over-70-year-olds. Numerous aetiological factors behind intellectual disability may explain this phenomenon. Yet, dementia, its risk factors and causes should be recognized and treated as early as possible also in the intellectual disability population. Further studies are needed to find the optimal healthcare management for this vulnerable group of citizens.

ACKNOWLEDGEMENTS
We would like to express our thanks to Tiina Häyrynen, Anne Keisu, Ulla Arvio and Anu Sorvisto, the nurse researchers, who collected the data for this study and Hannu Kautiainen for statistical analysis.

CONFLICT OF INTEREST
The authors declare no conflict of interest.

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How to cite this article: Arvio M, Bjelogrlic-Laakso N. Screening of dementia indicating signs in adults with intellectual disabilities. J Appl Res Intell Disabil. 2021;00:1–5. https://doi.org/10.1111/jar.12888