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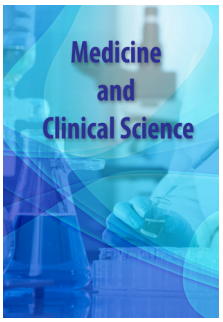
Review Article



Insights of Dementia in Persons With Intellectual Disability

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Abstract

Intellectual disability and dementia are age-dependent terms for a cognitive impair occurring during developmental age and in adulthood, respectively. Intellectually disabled people like any other people may develop dementia in adulthood. Thus, clinicians should learn to differentiate dementia-indicating signs from intellectual disability related cognitive deficiencies for an early diagnosis and treatment onwards. In intellectual disability, intellectual and adaptive skills of an individual are two standard deviations below the expected age-matched population, and dementia is characterized by a progressive cognitive decline. The cause of both disorders can be genetic, acquired or multifactorial. An increased risk of Alzheimer's disease in Down syndrome is well known unlike the development of dementia in other intellectual disability syndromes. This commentary discusses 1) how the dementia indicating signs present in intellectually disabled persons, 2) why it is important to distinguish dementia (and its causes) from intellectual disability and 3) why it is important to know the etiology of intellectual disability.

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Keywords

Intellectual disability, dementia, genetic, etiologies, differential diagnostics.

Introduction

The societal changes, improvements in living conditions and better education may decrease the occurrence of dementia in general population [1] and these factors can explain an extended lifespan of intellectually disabled persons [2]. Consequently, it is important to acknowledge the age-dependent health concerns such as dementia in health care management of also this vulnerable population [3]. Combination of the epidemiological and translational neuroscientific research is expected to result in better understanding of the various neuropathology underlying dementia disorders [1]. For instance, stress, diet and environmental factors known to regulate the anti-aging gene Sirtuin 1 may have a role not only in the aging process but also in the induction of intellectual disability and dementia [4-6]. We have approached this puzzle by studying the signs indicating dementia in people with intellectual disability of varying causes [2,7-10]. Such approach requires multidisciplinary teamwork of special units for the management of neuropsychiatric disorders of this patient group [11].

Intellectual disability vs. dementia

Intellectual disability and dementia are two age-dependent entities distinguishable from each other even though they share similar etiologies and symptoms [3,10,12]. Both intellectual disability and dementia result from dysfunctions of the cerebral cortex with overlapping etiologies i.e., genetic, acquired or multifactorial, however the causes of these two entities are not necessarily the same when they occur in the same individual. In intellectual disability the intellectual and adaptive skills of a child or adolescent are two standard deviations below what is expected for the age-matched group. Dementia is characterized by a progressive cognitive decline affecting the adult's ability to function independently. Both disorders impair many domains of the affected individuals such as the social, communication, cognitive, adaptive and motor skills.

Intellectual disability is recognized during the developmental age and its severity ranges from mild to severe or profound. The classification is based on intelligence quotient (IQ) testing, adaptive behavior assessment and whenever deemed necessary on other psychometric tests [9,13-15]. Intelligence quotient scores of 55 to 69 points refer to mild, 40-54 points to moderate and <40 points to severe or profound intellectual disability. In Finland, the assessment of intellectual disability severity done in adolescence determines the individual's capacity for further education, and that is also when an individual's cognitive development is considered to have reached a plateau (or at least slowed down). Consequently, the test result at adolescence has been used in our long-term follow-up studies as a baseline for later testing [8-10].

The cognitive decline that occurs in adulthood means the development of dementia, which proceeds from mild to moderate and

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from moderate to severe disease stages [16]. The more severe the stage of disease becomes the less active the affected persons become and the more simple activities (e.g. from complex arts and crafts to simple physical exercises) they end up doing in their everyday living [17]. Although dementia may develop at any age after developmental years, its risk usually increases along aging and its course is highly variable both between and within individuals depending also on dementia type [18].

Dementia indicating signs in intellectual disability

According to our own studies [9,10] the risk of dementia does not increase in the intellectually disabled population like in the general population along aging. When persons with ID of any etiology were screened, the dementia signs occurred as frequently in all age groups of adults [10]. However, when the dementia signs are evaluated separately in specific intellectual disability syndromes, the differences are evident as shown by our long-term follow-up studies [8,9,14,15]. Down persons start suffering from signs indicating dementia already soon after the age of 35 [8] and the risk of dementia in Williams's syndrome is similar as in general population [9,14] whereas men with Fragile-X syndrome do not appear to develop dementia at all [6]. Interestingly, however, cognitive aging in men with Fragile-X syndrome was found to start earlier (already before middle age) than in men in general population [15] and the course of cognition was found to be uneven in William's syndrome [14].

Dementia symptoms vary depending on the type of dementia and on the level of the individual's original intellectual capacity. The signs of dementia in intellectually disabled persons are screened by the British Present Psychiatric State-Learning Disabilities assessment [19,20]. This scale (incl. 27 items) has been used also in our studies [2,8-10]. In the study group consisting of 230 persons with intellectual disability of any cause, the most common dementia symptoms were reduced self-care skills, loss of energy, diurnal mood variation, forgetfulness and forgetting people's names [10]. The same most frequent first signs were noted also in 43 Down syndrome cases, who were screened three times during a 15-year follow-up [8].

One intriguing question is how dementia develops in different severity groups of intellectual disability. According to the British Present Psychiatric State-Learning Disabilities assessment, the severity of intellectual disability (as estimated in adolescence) did not predict the development of dementia in 138 study subjects consisting of pooled data from 62, 22 and 44 cases with Down, Williams and Fragile-X syndrome [9]. Nevertheless, the most common signs of dementia differed to some extent between the severity groups, ranging from subnormal to profound intellectual disability. The most common dementia signs among the three subjects with a profound intellectual disability were weight change, loss of energy and sleep disorders whereas the most common dementia signs of the four study subjects with mild intellectual disability were irritability, worry, weight change, and reduced self-help skills.

Evaluation of underlying causes of dementia signs in intellectually disabled patients

Thorough clinical examination and laboratory testing, we may reveal numerous different and clinically significant causes behind the signs of dementia in intellectually disabled patients. Right after the first dementia signs are noted one should exclude the conventional somatic, psychiatric or psychosocial factors that may have a negative impact on the patient's cognition.

Sometimes obtaining the right diagnosis requires further etiologic exams such as EEG-recording or genetic testing [7,21]. They may reveal unusual yet treatable disorders. For example, in one case dementia-like symptoms in a middle-aged man were found to be due to a childhood epileptic encephalopathy (CSWS, continuous-spike-wave in slow sleep syndrome, [7]). Such finding was not to be expected at such a late age, but once discovered it resulted in a full recovery following the initiation of a right epilepsy medication, i.e., clobazam. In another case, the re-evaluation of the etiology of intellectual disability revealed the Angelman syndrome, which helped to recognize the specific type of epilepsy, i.e., myoclonic status in non-progressive syndrome [18]. When the right medication, stiripentol, was initiated, the bed-ridden patient was literally able to leave her bed and walk. On the other hand, according to our clinical observations, a suboptimal medication may lead to an impaired intelligence in patients with epilepsy with eyelid myoclonia, whose cognition is usually normal or only borderline impaired [22]. Based on our clinical experience [7,21,22] we believe that continuation of a thorough evaluation of the underlying causes behind both intellectual disability and dementia can lead to new scientific discoveries.

Conclusion

Whenever any deterioration in a wellbeing of an intellectually disabled individual occurs, a clinician should consider the possibility of dementia or some other somatic disorder. Finding the cause behind dementia indicating symptoms may be laborious, but the right diagnosis at as early stage as possible is extremely important for the maintenance and restoration of intellectually disabled people's functional ability. Furthermore, the evaluation of specific etiology of both intellectual disability and dementia symptoms has turned out to be both scientifically interesting and clinically relevant. Based on our limited clinical experience quite unorthodox but treatable causes such as an unrecognized epilepsy type, unidentified genetic ID syndrome, or suboptimal epilepsy medication may be found behind dementia like symptoms or cognitive decline in intellectually disabled people.

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