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Neuropsychological profile of aging in mild intellectual disability: A case report

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ABSTRACT

People with intellectual disabilities (ID) are living longer, bringing into focus the need to better understand the cognitive trajectories associated with normal and abnormal aging in this population. The following case illustrates a neuropsychological profile of a 67-year-old Caucasian male, “Mr. Rodgers.” He has a diagnostic history of depression and anxiety. He is adopted as a child and much of his developmental history is unknown. The assessment follows a flexible battery approach and reveals a mild ID and a complex psychiatric profile associated with severe psychosocial trauma. He is treated with sertraline injections and individual and milieu therapy. Conclusion: The assessment of aging in geriatrics with ID requires the consideration of many factors that remain sensitive to pre-existing deficits and functioning. Norms stratified for age, education, and ID severity may improve the differential diagnosis of dementia in this population. The Neuropsychological Assessment Battery (NAB) shows promising utility and may be suitable when appropriate education estimations are used. Keywords: case report, mild intellectual disability, aging, memory, maltreatment.

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Introduction

According to the National Down Syndrome Society [1], 30% of people with Down's Syndrome (DS) in their 50's has Alzheimer's dementia (AD) with 50 % having the disorder in their 60's. Due to the advances in health care, individuals with intellectual disabilities (ID) are living longer. Despite this, age of death and mortality rates for individuals with ID even without comorbid developmental disabilities continue to demonstrate disparity [12]. In a recent study investigating the cause of death in individuals with ID in the United States, it was found the difference in age at death between adults with and without ID varied by presence of ID as well as ID severity. When compared to individuals without ID, greater variation in age of death was associated with severe/profound forms of ID (16.8 years), and less variation (10.3 years) was associated with mild/moderate forms [12]. The authors also found that when compared to adults without ID, individuals with ID had lower rates of death from heart disease, malignant neoplasms, cerebrovascular disease, chronic lower respiratory diseases, and dementia/Alzheimer's. Age is the greatest risk factor for the development of Alzheimer's dementia, suggesting that as the population aged 65 years and older continues to increase, the population impacted by dementia will continue to grow. Because individuals with ID without DS are living longer lives, it brings into light the question of the increased risk of the development of dementia and the need to clarify assessment approaches, definitions, and treatment recommendations.

Intellectual disability is diagnosed when an individual's intellectual quotient (IQ) is at least two standard deviations below population norms (IQ of 70 or below), with a comorbid deficit in adaptive functioning across conceptual, social, and practical domains [2]. According to National Center on Birth Defects and Developmental Disabilities[24], the cause of ID is usually unknown but can result from pre-natal, peri-natal, or post-natal injury, disease, or problem with the brain. The most common forms of ID develop from factors such as infection, genetics, disease, or teratogen exposure before birth. Other causes occur from stroke, head injury, or infection and can cause ID when an individual is older. Individuals with ID often perform much lower on neuropsychological measures than do the general population, making the detection of decline from baseline harder to ascertain. There is also an increased risk of depression and anxiety in later life in this population due to poor social adjustment, further victimization, and poor vocational outcomes [5,6,16,17]. Psychiatric and socio-cultural factors can challenge the detection of normal and abnormal aging patterns in the ID without DS population in later life, especially when considering pseudodementia against mild cognitive impairment (MCI) and/or dementia. What remains unclear is the relative risk of developing dementia in the ID without DS population. Previous studies have attempted to estimate the relative risk of developing MCI and dementia in individuals with ID, but results have been inconsistent [23,29]. Even more, the definitions used to classify cognitive impairment in this population are unclear and could account for variability in study outcomes [10].

Timeline

Table 1 [*appendix*] demonstrates the timeline of events leading up to Mr. Rodgers' referral to neuropsychology.

Narrative

Presenting Problem

Mr. Rogers reports that his problems with word-finding, misplacing items, and forgetting recent events have been onset over the last year. He complains of processing information slowly, poor concentration, and inability to perform mental calculations. He reports insomnia and occasional headaches. Mr. Rodgers can bathe, use a toilet, walk, and dress himself independently. He can still perform yardwork/housework, use a telephone, manage medication, and grocery shop. He struggles to managing finances/pay bills and prepare meals. He explains that recently while cooking, he used

a glass bowl to boil water, which exploded in shards. His adoptive sister sates that Mr. Rodger's has "always" had some problems with complex multi-step tasks, memory, and thinking, but has never been diagnosed with an intellectual disability. Emotionally, Mr. Rodgers expresses a significant interpersonal resentment. He complains that people manipulate and take advantage of him. He has chronic feelings of worthlessness and low self-esteem as well as problems with anger and depression. He struggles to trust others and describes himself as chronically lonely. At times throughout the interview, he fixates on death and suicide fantasies. He denies having any friends, and reportedly "fears women."

Mr. Rodgers lives in 4 foster homes before he is adopted at the age of 15. Although his adoptive family is warm and supportive, he struggles to adjust socially outside of the orphanage. Added stressors include living alone, financial burden, and stress about his health functioning. His adoptive sister joins via ZOOM to provide historical and collateral information.

Social & Developmental History

As a child, Mr. Rodgers suffered a severe beating reportedly perpetrated by his biological parents. He is placed in the NICU for a non-disclosed amount of time, and states he does not kick a ball or use a pencil until the age of 5. He does not read or write until the age of 8. Accordingly, a 15-year-old girl at the orphanage, who Mr. Rodgers refers to as "a mommy," taught him how to read and write. He transitioned to public school in the 8th grade but is “never” able to pass tests or grasp the material. He gets mostly D's, and states he was “passed through” until the 11 grade. He eventually discontinues formal education at the age of 17 and does not complete a GED program. He denies current addiction or abuse of alcohol, recreational drugs, or nicotine. As a child he is bullied, with other children referring to him as "dumb." He adds that as an adult coworkers become frustrated with him and call him "idiot." He has 1 previous marriage that lasts two years, ending due to infidelity of his reported exploitation of his wife.

Adaptive Functioning

Mr. Rodgers struggles with independent living throughout his life due to problems with managing finances and social adjustment; he moves-in with his parents on several occasions as a young and middle-aged adult. His adoptive parents passed away several years ago and he remains in close regular contact with his adoptive sister, who lives several states away. His sister helps him make important health and financial decisions. Mr. Rodgers worked for his father’s company on- and-off for 15 years and eventually receives his CDL license at the age of 57. After 5 years, his license is revoked when he fails to hitch his trailer properly, resulting in an accident. Due to previous car repossession and his motor vehicle accident, Mr. Rodgers no longer drives. Instead, he uses a bike with GPS to navigate the community. He denies any accidents or episodes of getting lost but relies heavily upon GPS.

Behavioral Observations

Mr. Rodgers struggles to complete a written history packet questionnaire on his own, leaving many items blank. He takes an equal role in the neurobehavioral interview and answers questions, at times referencing his adoptive sister to confirm autobiographical information and discuss reported symptoms. He is oriented to person, place, and situation. Prosody and speech volume are normal. His rate of speech is slow with childlike syntax and tone. Throughout testing, he requests words to be defined. He is verbose in speech, requiring consistent redirection. Mr. Rodgers is non- responsive to non-verbal cues in conversation. Interpersonally he is warm and friendly with staff and can engage in “story telling” with others. On testing days, he arrives several hours early, as he uses a transportation service. He is hungry at all sessions and does not eat breakfast. He also reports insomnia. He exhibits significant emotional reactivity such as crying and cursing throughout the testing session during challenging tasks. At times he refers to himself

as an “idiot.” There is no evidence of hallucinations or delusional thinking. His affect is anxious with congruent thought content. He ambulates unassisted with posture with his head slightly too far forward with shuffling feet, and slouched posture. He endorses chronic suicidal ideation without current plan or intent. He exhibits impaired attention and concentration. His autobiographical memory is below average, but he exhibits normal recent episodic memory. He can mull over and worry about his test performance information in between sessions. Audition is normal. Vision is impaired but reaches normal thresholds with use of corrective lenses.

MRI of brain without contrast 12/02/2021

MRI of the brain dated 12/02/2021 indicates no evidence of cerebral infarct, white matter disease, hematoma, mass detection, hydrocephalus, or extra-axial fluid collection. There is no midline shift and cerebral flow appears normal. Overall impression indicates a normal examination without remarkable evidence.

Psychiatric Treatment

Mr. Rodgers reports receiving psychotherapy for his psychiatric diagnoses as a youth. To manage his current mood disorders, he receives care at an outpatient psychiatric clinic and is treated with Sertraline injections (60 mg PO QHS).

Methods

Comprehensive Testing Battery

Table 2 [*appendix]* indicates the psychometric measures administered. The presented *T* scores are based on United States norms based on age, sex, and education wherever possible. Generally, percentile scores at or below the 1st percentile ($T \leq 28$) were considered impaired, scores ranging from the 2nd to 8th percentile ($29 \geq T \leq 36$) were considered borderline, scores ranging from the 9th to 24th percentile were considered low average ($37 \geq T \leq 42$), and scores ranging from the 25th to 74th percentile ($43 \geq T \leq 56$) were considered average. Standard Scores have a mean of 100 and a standard deviation of 15. Scaled scores have a mean of 10 and a standard deviation of 3. BDI-II Score Range: Severe 29-63 Moderate 20-28 Mild 14-19 Minimal 0-13; BAI Score Range: Severe 26 – 63, Moderate 16 – 25, Mild 8 – 15, Minimal 0 – 7.

Transparency and Openness

This study presents citations in standard with guidelines for transparency and openness promotion (TOP) in research. De-identified raw data is available upon request to the author. Study materials can be purchased from test developers to individuals with qualifying credentials. No aspect of this work is preregistered. Written informed consent to publish the case details was obtained from the patient and his adoptive sister.

Discussion

Intellectual Functioning

Results of the current evaluation are consistent with a diagnosis of mild intellectual disability with a RIAS 2 composite intellectual quotient of 64. Across verbal and non-verbal modalities of inductive reasoning, Mr. Rodgers' performance is unitary in the Extremely Low range of functioning. His premorbid IQ is expected to fall somewhere in the Extremely Low to Borderline range of functioning, consistent with his current IQ scores. He does exhibit a personal strength in deductive verbal reasoning (RIAS 2 "Guess What" subtest), a form of deductive reasoning which is required in typical social conversation. Mr. Rodgers' relative strength in deductive verbal reasoning and social communication effectively masks deficits which may result in others’ estimation of his reasoning ability and comprehension to be higher than it is. This may even help explain why his ID went undetected his entire life. There is good inter-test reliability between his RIAS 2 and WAIS-IV scores, however, the WAIS-IV verbal subtests require a respondent to induct verbal information and produce a response, whereas the 'Guess What' subtest of the RIAS2 requires deductive verbal constructs. This key structural difference between the WAIS-IV and RIAS2 may explain Mr. Rogers’ cognitive test variability, where

context clues help him to deduce correct answers.

Although Mr. Rodgers was first administered the WAIS-IV, he is ultimately unable to complete the test due to emotional reactivity and frustration. Nevertheless, his WAIS-IV verbal comprehension ability (SS = 68), thought to measure crystallized intelligence, reflects his low academic achievement and is minimally discrepant from his RIAS2 composite score. The WAIS-IV matrix reasoning subtest, which measures inductive nonverbal abstract reasoning can also predict academic success as a factor of fluid intelligence [25]. Since Mr. Rogers' WAIS-IV matrix reasoning subtest performance (ss = 5) suggests moderate impairment in fluid and inductive reasoning, he is likely to struggle in nonverbal problem-solving and complex applications, such as mathematics, explaining his lifelong difficulty with financial management and decision-making. Lastly, WAIS-IV trials on special populations, including mild and moderate ID suggests ID populations demonstrate a tendency for subtest performance to fall in the 3 – 5 range for scaled scores [27]. This is consistent with the Mr. Rogers' overall WAIS-IV subtest performance, suggesting positive confirmation of mild intellectual disability and performance consistent with his global RIAS2 score.

Executive Functioning

Mr. Rogers' dysexecutive function is non-unitary. On one hand, he exhibits areas of executive deficit involving self-monitoring, mental flexibility, emotional control, inhibition, and abstraction. Moreover, low frustration tolerance and a propensity toward emotionality in the face of a challenge further impairs complex problem-solving. On the other hand, Mr. Rodgers' judgment, planning, and organization meet normal thresholds when adjusted for age and education level (8 years) using the Neuropsychological Assessment Battery (NAB). These findings demonstrate the impact of limited education on his executive functioning and may also suggest the suitability of the NAB for use in the ID population. The heterogeneity of ID and the presence of underlying executive deficits increases the difficulty of establishing pre-morbid functioning [10]. Mr. Rodgers was not provided a measure of pre-morbid functioning, as assessment tools that estimate premorbid functioning may be irrelevant in the ID population and should not be considered unless estimations are derived from a comparative norming sample.

His phonemic fluency is consistent with his performance on verbal measures of the WAIS-IV and is regarded as normal for his intellectual functioning. However, on the RIAS2 'What's Missing' subtest, which measures visual attention, gestalt formation, concept formation, and deductive reasoning, he achieves a low score. 'What's Missing' is a complex executive task that "loads" on executive functioning and attentional processing, thereby decreasing the efficiency of his non-verbal reasoning ability. Changes in functional ability related to Alzheimer's disease may be subtle for individuals with ID. Mr. Rodgers had always struggled to complete IADLs. Therefore, the presence of deficits in IADLs alone is not enough to justify neurodegeneration in the direction of MCI or dementia. Because capturing relative decline in adaptive functioning for ID populations is difficult, caregiver and family collateral input as well as record reviews must be integral to the assessment process.

Language & Memory

It is believed that dementia in individuals with ID displays a conventional pattern of decline in memory functioning presenting as an amnestic profile. However, the heterogeneity of ID makes it difficult to decipher if noted areas of deficit are due to natural stratification related to ID pathology, or if they constitute a change from baseline. For individuals with ID and DS will experience early signs of alterations in episodic memory [11], whereas individuals with ID without DS may present first symptoms of aging as behavioral changes. While Mr. Rodgers' memory profile is variable, his pattern of deficits is not consistent with typical neurocognitive decline. There is no anomia or rapid forgetting

and neurodiagnostic imaging is unremarkable. He obtains a 12/15 on a confrontation naming test, missing only two items he has never heard of. Because performance on the Boston Naming Test (BNT) can be a function of IQ and language ability, errors are likely related to his low educational achievement [15].

Mr. Rogers' memory scores range from moderately impaired to low average. Fluctuation in memory is often associated with fluctuations in cognitive efficiency, which can suggest the presence of mood disorders impacting concentration, memory, and attention [30]. At the time of the evaluation Mr. Rodgers reports severe depression (BDI-II Total = 54) and moderate anxiety (BAI total = 24), which likely contributes to test performance variability. However, overall, his composite memory scores are consistent with his intellectual ability and thus, are interpreted as normal and consistent with his baseline functioning. Low baseline functioning and "cognitive reserve," in individuals with ID may exaggerate deficits as part of a normal aging process, potentially leading to false positive detections of dementia when using standard neuropsychological measures.

Psychological Functioning

Individuals with ID are at increased risk of suicide [26] and children with ID are more likely to experience physical abuse than children ID [21,22]. Maltreatment of children that results in traumatic brain injury has been linked to other aversive outcomes such as increased risk of future violence and victimization, learning difficulties, difficulty finding employment and succeeding in school, cognitive problems, and delayed brain development [5,6,17]. It is unclear if Mr. Rodgers' ID is congenital or acquired, and whether mode of contraction of ID is associated with dementia risk.

Mr. Rogers fixates on a need for "a mommy," which may suggest an early psychodynamic trauma which may contribute to his "fear of women" and/or history of exploitation by romantic partners. Traumas related to social determinants of health (SDOH) can lead to complex clinical presentations involving physical, neurological, and psychological changes. ID populations are at greater risk of aversive life experiences, which may impact AD disease course and expression. Furthermore, Mr. Rodgers experiences neglect during critical periods of development, which may contribute to aspects of his cognitive functioning. Further investigation of this SDOH and AD risk in individuals with ID is warranted.

Self-Report measures

There is concern about using self-report measures for individuals with ID, as getting access to valid and reliable information can be a struggle due to problems with comprehension and insight. This appears in the case of Mr. Rodgers, where he notes cognitive problems onset in the last year, but his sister reports cognitive problems have been lifelong. To address deficits in introspection and insight, researchers consider one of three options: (1) use measures that can be administered without alternation in form, (2) use measures that can be administered after making adaptations, and (c) use measures specifically designed for individuals with ID [9]. It is common practice to adapt measures, and many measures have been adapted [3,7,13]. Criticism of adaptations includes the lack of robust reasoning for the specific adaptations that account for *all* possible challenges associated with self-report assessment in this population. There are also measures that are specifically designed to account for the needs of people with ID, such as the Glasgow Anxiety Scale for people with Intellectual Disability [14] and the Lancaster and Northgate Trauma Scales [28]. Previous systematic reviews have reviewed the effectiveness of various instruments for diagnosing dementia in adults with intellectual disabilities [4]. However, these measures are not routinely part of a neuropsychologist's compendium of tests. The heterogeneity of the ID population and the absence of comprehensive and practical evidence-based guidance for practitioners who use suggested measures with ID populations may contribute to

misdiagnosis and mistreatment.

Patient Perspective

Mr. Rodgers struggles to articulate his desired outcome at the time of testing but does report significant frustration about managing finances, living alone, and being able to problem-solve. Throughout the testing process, he thanked the staff and providers for their kindness and reports general satisfaction with the interpersonal aspect of his health visits. After receiving feedback, he expresses relief and reports feeling validated and with newfound direction. His adoptive sister shares in his sentiment. Mr. Rodgers was contacted 5 months after his visit and provides an update. He is working with a case manager to help navigate social security and adult living options. He continues with psychiatric and psychotherapeutic treatment, and his depression has improved. He still expresses some frustration with navigating interpersonal contexts and has made steps toward establishing an appointed power of attorney and moving into an adult assisted living community.

Conclusion

Pre-existing deficits in memory, processing, working memory, attention, and executive functions are associated with mild to severe forms of ID. The presence of pre-existing deficits can make it difficult to differentiate normal aging from abnormal aging in this population. Identifying cognitive trajectories in the ID populations is not straightforward and requires a thorough history and consideration of demographically corrected norms for age and education to understand functioning. The Neuropsychological Assessment Battery (NAB) showed promising support for use in the ID population when correct education adjustments were made. Relying heavily on the patient's report for history may prove problematic, as insight into individuals with intellectual disability may be impaired.

The current compendium of neuropsychological tests was not developed to differentiate those with ID from those with cognitive impairment, where in cases of ID, only a small amount of additional neuropathology is needed to impact functioning beyond the baseline. The results of the current case report have limited generalizability due to the heterogeneity of ID, underscoring the importance of establishing sensitive tools to improve dementia diagnosis in this population. A flexible battery approach to neuropsychological assessment is recommended to better capture functioning as it relates to areas of strength and weakness. A health systems approach, using integrated health systems in psychiatry, neuropsychology, psychotherapy, and social supports is recommended to improve treatment outcomes.

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Conflict of interest

There are no conflict of interests to report.

Ethical approval:

Not applicable

Consent for publication

Written and verbal informed consent to publish deidentified case details was obtained from the patient and his adoptive sister.

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Appendix

Table 1 – Timeline

2018-01-30	Mr. Rodgers has a MVA with no resulting injury. His license is revoked.
2018-03-29	Mr. Rodgers "loans" his car to a girlfriend. Payments go into default and his car is repossessed.
2021-11-09	His experiences problems with word-finding.
2022-01-03	Homeless individuals move-in with Mr. Rodgers in exchange for home health services. An adult protective service case is opened.
2022-03-21	Mr. Rodgers' attempts to boil water with a glass bowl, resulting in a cooking accident without injury to himself or others.

Table 2 Intellectual Functioning

Intellectual Functioning	
Weschler Adult Intelligence Scale-Fourth Edition (WAIS-IV)	
Verbal Comprehension	SS = 68
Similarities	ss = 4
Vocabulary	ss = 5
Information	ss = 4
Perceptual Reasoning	
Matrix Reasoning	ss = 5
Processing Speed	SS - 65
Symbol Search	ss = 3
Coding	ss = 4
Working Memory	
Digit Span	ss = 3
Longest digit span length	ss = 4

Reynold's Intellectual Assessment Scale-Second Edition (RIAS-2)	
Verbal Intelligence	SS = 81
Guess What	T = 42
Verbal Reasoning	T =33
Nonverbal Intelligence	SS = 55
Odd Item Out	T = 34
What's Missing	T = 9
Composite Intellectual	SS = 64

Learning and Memory

Learning and Memory	

Weschler Memory Scale - Fourth Edition (WMS-IV)	
Logical Memory I	ss = 3
Logical Memory II	ss = 4
Visual Reproduction	ss = 5
Visual Reproduction II	ss = 7
California Verbal Learning Test-Third Edition (CVLT-3)	
Immediate recall	SS = 63
Trial 1 (raw score)	3
Trial 2 (raw score)	4
Trial 3 (raw score)	4
Trial 4 (raw score)	4
Trial 5 (raw score)	3
Delayed Recall	SS = 65

Executive Functioning

Executive Functioning	
Delis Kaplan Executive Functioning System	
Color-Word Interference	
Color Naming	ss = 1
Word Reading	ss = 1
Inhibition	ss = 1
Inhibition/Switching	ss = 1
Trail Making Test	
Trail Making Condition 1	ss = 1
Trail Making Condition 2	ss = 1
Trail Making Condition 3	ss = 1
Trail Making Condition 4	ss = 1
Trail Making Condition 5	ss = 7
Verbal Fluency Test	
Letter Fluency Test	ss = 3
Category Fluency	ss = 4
Category Switching	ss = 2
Total switching accuracy	ss = 7
Neuropsychological Assessment Battery *(NAB)	*Using 8th grade education adjustments
Executive Functioning	T = 50

Mazes	T = 51
Judgement	T = 56
Categories	T = 57
Word Generation	T = 40
Clock Drawing	WNL (9/10)

Adaptive Functioning

Independent Living Scale (ILS)	
Money Management	T = 20

Language

Language	
Boston Naming Test (BNT) Short Form	12 of 15 Patient does not name sphinx and palette.
Repetition	Intact for simple and complex sentences
Receptive and expressive language	He understands idioms and colloquial language use. He often engages with examiner using humor, demonstrating reciprocal and spontaneous discourse, as well as logical and goal-oriented speech.

Emotional Functioning

Emotional Functioning	
Beck Depression Inventory - Second Edition (BDI-II)	Total score = 54 [1] BDI-II Score Range: Severe 29-63 Moderate 20-28 Mild 14-19 Minimal 0-13
Beck Anxiety Inventory (BAI)	Total score = 24 BAI Score Range: Severe 26 – 63, Moderate 16 – 25, Mild 8 – 15, Minimal 0 – 7
Rotter's Incomplete Sentences Blank	Men…can't stand; Most women…abuse me, use me, lie to me; I hate…mean people; Other people…no manners; Marriage…never; What annoys me…people; People…not honest. (social problems/traumas) I need…a mommy; A mother…I love mommy's but never had one; My father…never had one, I hate them (psychosocial trauma in object relations) I can't…do nothing; My mind…can't remember, useless; I am very…different; I…am not (awareness of intellectual deficits, negative self-concept/ self-efficacy beliefs)