

## APPENDIX G

# Dementia and Adults with Intellectual / Developmental Disabilities

*Nancy Jokinen, MSW, PhD, Associate Professor, BSW Program Coordinator, University of Northern British Columbia, School of Social Work*

Adults with intellectual disabilities (ID) are living longer with some individuals reaching ages seen in the general population (Coppus, 2013; Patja, Molsa, & Iivanainen, 2001). Dementia as it affects adults with ID is a growing significant concern for families, services and policy-makers. The World Health Organization and Alzheimer's Disease International (2012) recognize people with intellectual disabilities and their caregivers as having unique needs (p. 54). Additionally, an International Summit on Intellectual Disabilities and Dementia (Glasgow, Scotland – October 13-14, 2016) brought together leading academics and organizational representatives from across Europe and North America. Their work produced a number of summative reports on pressing issues including inclusion in national plans, nomenclature, end-of-life care in advanced dementia, and post diagnostic supports (for a full listing see: National Task Group on Intellectual Disabilities and Dementia Practices, 2018). Various research projects, predominately from other countries, have also focused on dementia related to this often-marginalized group. A sampling of recent studies includes:

- An English study examining diagnosis, survival and risk factors associated with Alzheimer's disease and Down syndrome (DS) utilizing the Aging with Down Syndrome and Intellectual Disabilities (ADSID) research database (Sinai *et al.*, 2018).
- An Irish longitudinal study following adults with DS examined age of onset, dementia features, and co-morbidities (McCarron *et al.*, 2017).

- A United States study investigating aging, dementia and multimorbidity in relation to adults with Down syndrome aged 45 – 89 years (Bayen, *et al.*, 2018, July 22).
- A longitudinal study in the United States follows three dementia-care community-based group homes to observe progression of decline, resident needs, and practice adaptations (Janicki, 2018, June 29).
- The INCLUDE (INvestigation of Co-occurring conditions across the Lifespan to Understand Down syndromeE) project (National Institutes of Health, 2018).
- Studies by the Alzheimer's Biomarkers Consortium — Down Syndrome (ABC-DS) (2018).

Within a Canadian context, however, few research projects report on dementia as it affects adults with ID. As a result, there is a reliance on international research to inform Canadian policies and practices despite differences in healthcare and social service systems. Additionally, across Canada, the provinces and territories differ in their respective health and social services as well as data collection strategies in relation to this population, making it difficult to compile national evidence on dementia affecting adults with ID and or DS. A cursory review of provincial strategies found no specific actions related to individuals with ID and or DS and their families.

This paper highlights extant evidence pertaining to adults with ID affected by dementia including prevalence of dementia, assessment and diagnosis, and family caregivers. It draws heavily upon international work. The inclusion of adults with ID and their families in national plans developed by other countries is then considered.

## PREVALENCE OF DEMENTIA

Studies examining the prevalence of dementia affecting adults with ID without Down syndrome (DS) report mixed findings. In the United States, Janicki and Dalton (2000) found an overall prevalence of 6.1% for adults aged 60+, similar to rates in the general population. Zigman *et al.*, (2004) also found comparable rates of dementia. Yet other studies suggest higher prevalence. For example, Cooper (1997) found a prevalence of 21.6% in the UK, much higher than expected for a comparable age group in the general population. In Manitoba, Canada, a prevalence of 13.76% was reported for adults with ID aged 55+, again higher than prevalence in the general population (Shooshtari, Martens, Burchill, Dik, & Naghipur, 2011). Methodological differences including criteria used to diagnose dementia likely explain these differences in the research findings (Silverman, Zigman, Krinsky-McHale, Ryan, & Schupf, 2013; Strydom, Livingston, King, & Hassiotis, 2007).

That said, there is longstanding agreement that adults with DS are at high risk of acquiring dementia. Generally, estimates suggest 25% will be affected after age 40 and at least 50 to 70% are affected after age 60 (National Task Group on Intellectual Disabilities and Dementia Practice, 2012). McCarron, *et al.* (2017) confirmed a high risk associated with Down syndrome that ranged from 23% for individuals aged 50 years to 80% for adults aged 65+. Sinai, *et al.* (2018), also reported a younger average age at diagnosis (55.80 years) and a reduced survival time from diagnosis (average 3.78 years) for people with DS. Bayen, *et al.* (2018) also confirmed a high rate of dementia in a California study of people with DS as well as an increased number of comorbid conditions in those adults affected by dementia compared to those without dementia, particularly hypothyroidism, epilepsy, anemia and weight loss.

## ASSESSMENT AND DIAGNOSIS

Guidelines for the assessment and management of dementia as it affects persons with ID were first published in the mid 1990's, see Janicki, Heller, Seltzer and Hogg (1996). These alongside other reports including community support guidelines (Jokinen *et al.*, 2013) and an assessment framework for physicians (Moran, Rafii, Keller, Singh, & Janicki, 2013) recognize the challenges inherent in assessment and diagnosis of dementia for people with ID. Canadian primary care guidelines also offer recommendations for action in regards to assessment of dementia (Sullivan, *et al.*, 2018).

Assessment requires the use of instruments different from those used with the general population and Table 1 offers examples of commonly used measures for clinical assessment. Best practice guidelines do speak to the need for healthcare professionals to be familiar with the population to avoid diagnostic overshadowing. The assessment process critically compares previous with changed behavior and function. The guidelines all suggest adults with an ID have an established baseline of abilities to facilitate the assessment process if needed. There are a number of options to record such a baseline (e.g., video, pen/paper). The NTG-EDSD (Esralew *et al.*, 2013; National Task Group on Intellectual Disabilities and Dementia Care Practices, 2013) may also be used for this purpose (Jokinen, *et al.*, 2013). This is, an administrative tool specifically designed for family and direct care staff to document change over time and discuss any changes noted with a healthcare professional. Its usefulness has been noted in a German study (See Zeilinger, Gärtner, Janicki, Esralew, & Weber, 2016) and the tool has been adopted in a number of jurisdictions. In North America, a baseline of abilities has been recommended for people with DS commonly beginning at age 30 or 40 or 30 and by age 40 or 50 for individuals with other ID to facilitate assessment should the need arise.

**Table 1: Measures for Clinical Assessment of Dementia in People with Intellectual Disabilities\***

Name of Measure	Instrument characteristic	Strengths/weaknesses
Adaptive Behaviour Dementia Questionnaire (ABDQ) (Prasher, Farooq, & Holder, 2004)	15-item questionnaire used to detect change in adaptive behavior by comparing current functioning to typical functioning.	<p><b>Strengths:</b> Detecting change in everyday functioning, easy to administer and score, family member as informant, length of time must know adult is specified.</p> <p><b>Weaknesses:</b> No differential diagnosis, not useful to track intervention response.</p>
Assessment for Adults with Developmental Disabilities (AADS), (Kalsy, McQuillan, Oliver, & Hall, 2000); (Oliver, Kalsy, McQuillan, & Hall, 2011)	Measure of function with focus on behavior and performance related to cognitive and physical decline; 28 items with respect to 'how often', 'management difficulty', and 'effect'.	<p><b>Strengths:</b> Assesses current everyday functioning/ behavior, and behavior changes. Easy to administer, standardized administration, descriptive, indicates who needs further evaluation/care, many informants.</p> <p><b>Weaknesses:</b> No differential diagnosis. Questions are complex.</p>
Dementia Questionnaire for People with Learning Disabilities (DLD)*, (Evenhuis, 1992); (Evenhuis, 1996); (Eurlings, Evenhuis, & Kengen, 2006)  *Originally named the Dementia Questionnaire for Mentally Retarded Persons (DMR)	Made up of eight sub-scales: short term memory, long term memory, orientation (making up Sum of Cognitive Scores), speech, practical skills, mood, activity and interest and behavioral disturbance (making up a Sum of Social Scores).	<p><b>Strengths:</b> Designed for all levels of functioning as early screening instrument. Easy to administer as informant completion item or interview. Indication of dementia signs at one assessment and over repeated assessments.</p> <p><b>Weaknesses:</b> Level of functioning (e.g., IQ) required for norms. No differential diagnosis. Some concern among clinicians regarding its appropriateness for individuals in the severe and profound ranges of intellectual functioning.</p>
Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID), (Deb, Hare, Prior, & Bhaumik, 2007)	Comprised of 43 questions in three sections. Measures memories, confusion, feelings of insecurity, sleep problems, and behavior problems. Includes information about medical conditions, psychiatric conditions, and medication.	<p><b>Strengths:</b> Current everyday functioning assessed, easy to administer/score, wide range of respondents considered appropriate, length of time informant needs to know adult specified.</p> <p><b>Weaknesses:</b> For differential diagnosis just lists possible other conditions and medications.</p>

\* Adapted from Source Jokinen, *et al.* (2013, p. 7)

## FAMILY CAREGIVERS

Research and the literature point to the varied and unique situations of family caregivers faced with the prospects of dementia care (see as examples the narratives in Jokinen *et al.*, 2018). Many adults with ID continue to live with family in middle and older age. In the United States, for instance, 71% of adults live with family – 24% with parents aged 60+ (Braddock, Hemp, Tanis, Wu, & Haffer, 2017). Other middle-aged and older adults with ID live in varied circumstances (e.g., alone, with unrelated adults in home shares or group homes). Family members living with their relative with ID affected by dementia provide primary support and those living separately often remain involved in supportive roles (Jokinen, *et al.*, 2018). Many families remain committed to provide support despite changes encountered with dementia (Janicki, Zendell, & DeHaven, 2010).

Most adults with ID and their families have different life experiences than people in the general population affected by dementia. They have:

- decades long involvement with various education, health and social services
- long time exposure to stigmatization, discrimination, and exclusion
- and endured changes in philosophy and social support from institutional care to community living (Jokinen, 2016).

These family caregivers are a distinct group often being older-aged parents or siblings versus a spouse or adult offspring and providing care for decades since the birth of their relative with ID compared to assuming responsibilities later in life (Jokinen, *et al.*, 2018). After providing a lifetime of support, families face new challenges when dementia presents and begin to question their abilities to continue as caregivers given the transitions from established routine care to that related to stage-related changes (Heller, Scott, Janicki, & Pre-summit Workgroup on Caregiving and Intellectual/ Developmental Disabilities, 2018). The literature, however, is sparse on how families cope with the various transitions encountered in dementia care (Jokinen, Janicki, Hogan, & Force, 2012).

## INCLUSION OF ADULTS WITH ID AND THEIR FAMILIES IN NATIONAL PLANS

Despite the recognition of adults with ID as a unique population that should be included in national plans (World Health Organization & Alzheimer's Disease International, 2012), a review of national plans indicates a number of them make essentially a descriptive mention of adults with ID and their families and nominal attention is given to the challenges caregivers encounter with dementia. Yet other countries have made some specific mention including the latest iteration of Norway's Dementia 2020 plan, (Watchman *et al.*, 2017). Another example is the USA National plan that has included specific mention of adults with ID, possibly in part because of the work of the National Task Group on Intellectual Disabilities and Dementia Practices (NTG) and their frequent presentations at meetings of the Federal Advisory Council on Alzheimer's Research, Care, and Services (Janicki & Keller, 2014). The current update on the US Plan (U.S. Department of Health and Human Services, 2017) includes the following specific references in reference to ID / DS:

- Strategy 1.C: Accelerate Efforts to Identify Early and Presymptomatic Stages of Alzheimer's Disease and Related Dementias. Refers to the work of the National Institutes of Health's Alzheimer's Biomarker Consortium-Down Syndrome (p.15).
- Recommendation 1: The 2017 National Plan should continue to provide a robust, comprehensive, and transformative scientific Road Map for achieving the goal of preventing, effectively treating, and providing effective care and services for AD/ADRD by 2025. Speaks to establishing research milestones that "Include and prioritize specific milestones for populations at high risk for AD/ADRD (e.g., people with Down syndrome, African Americans)" (p.62).
- Recommendation 17: Federal agencies, states, national health and aging organizations, and community partners must continue to expand public awareness and training, reduce stigma, and help connect people to information and available resources. Outreach should include children and youth, diverse racial/ ethnic/socioeconomic groups, and people with IDD.

Includes: "Increase engagement of national health-related organizations (e.g., Down syndrome, heart, and diabetes) in providing information and resources addressing dementia" as well as "Beyond research-specific efforts, NIH is also committed to continuing to enhance and provide evidence-based information, resources, and referrals through the ADEAR Center to specific populations of people with AD/ADRD and their caregivers including younger people, non-traditional families, people with IDD, such as Down syndrome, and ..." (p. 75).

## CONCLUSION

Adults aging with ID and their family caregivers affected by dementia have unique experiences and needs compared to people in the general population. A lack of Canadian research and reliance on international research to guide policy and practice negates our healthcare and social context. A Canadian National Dementia Strategy that includes actions to address the needs of adults with ID and their families affected by dementia may provide an impetus for localized dementia strategies to also incorporate similar actions as well as begin to inform policy and practice for all persons affected by dementia.

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