

Editorial

Why do we need national guidelines for adults with intellectual disability and dementia?

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1. Introduction

The World Health Organization's (WHO) report, *Dementia: A Public Health Priority* [1] drew attention to the growing impact that Alzheimer's disease and related dementias are having on the world's older population. The report noted the need for more effective early detection and screening, diagnostic and medical treatment services, and innovative models of family supports and social care. It also identified "populations with specific needs," including adults with intellectual disability (ID)³ (and in particular adults with Down syndrome—who "are at a significant risk of developing Alzheimer's disease") as needing special attention. Others [4,5] have noted that such adults were at greater risk and had a differing course of dementia, often with earlier onset and shorter duration, and that they posed a diagnostic challenge for many practitioners. Also, the Edinburgh Working Group on Dementia Care Practices [6], an ad hoc international body, proposed that given these factors, service providers and national Alzheimer's organizations provide equity in services at an earlier age to people with ID.

The WHO report noted that dementia is often underdiagnosed and that this may be the result of three factors: stigma associated with dementia, beliefs that memory problems are a normal part of aging, and the belief that nothing can be done—these factors also all apply to the underdiagnosis of

dementia among adults with ID. The report proposed that to circumvent these factors, detection and diagnosis require coordination among health and social care systems and better attention to symptom presentation among primary and specialist care services. The WHO contended that initial identification is an important function not only of primary care, but also of the social care community which often has the best opportunity to have an impact when noticing behavioral and functional change. With respect to early identification, the WHO proposed that guidelines for practitioners and others involved with day-to-day dementia care "should include guidance on clinical aspects such as diagnosis, assessment and treatment, as well as quality long-term care" (p.62) [1].

Why is there a need for special guidelines for adults with ID? Individuals with ID are living longer including those with Down syndrome, and the risk of manifesting Alzheimer's disease and related dementias is increasing. Most families, community support organizations, and health

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³The World Health Organization [2] notes that *intellectual disability* is a condition with "a significantly reduced ability to understand new or complex information and to learn and apply new skills (impaired intelligence). This results in a reduced ability to cope independently (impaired social functioning), and begins before adulthood, with a lasting effect on development." In the United States, the National Task Group on Intellectual Disabilities and Dementia Practices [3] defined adults *with intellectual disability affected by dementia* as those who (1) have intellectual limitations that significantly limit the person's ability to successfully participate in normal day-to-day activities such as self-care, communication, work, or going to school, and (2) developed the intellectual limitation during the "developmental period" (before approximately 22 years of age), and (3) the limitation is anticipated to result in long-term adaptive or functional support needs, and/or (4) are eligible for State or Federal public support programs because they have been diagnosed as having an intellectual disability; and (5) are affected by dementia, and meet the criteria of having been diagnosed with possible, probable, or definitive dementia, or mild cognitive impairment, as defined by the World Health Organization's International Classification of Diseases or who meet the diagnostic criteria of the American Psychiatric Association's Diagnostic and Statistical Manual.

care practitioners are not familiar with how age-related decline may typically manifest, including pathologic decline from underlying disease. A change or decline in function may be falsely blamed on the underlying ID (i.e., diagnostic overshadowing) and many adults with ID may go undiagnosed or misdiagnosed. These uncertainties and lack of awareness often can create a delay in recognition that dementia is present, and therefore, appropriate evaluation, treatment, and supports may not be provided. Furthermore, generally accepted measures and procedures for diagnosis often are not appropriate for adults with ID because of their variable functional and intellectual premorbid state. Guidelines offering standardized procedures for screening, assessment, and diagnosis would mitigate many problems associated with accurate ascertainment of the presence of dementia.

Furthermore, guidelines for the care and support of adults with ID affected by dementia would go far to improve this growing personal, social, and health problem. With this in mind, the Edinburgh Working Group had noted that “in many jurisdictions, practices have been inconsistent with regard to how to provide services and supports to people with [intellectual disability] affected by dementia” and that “providers have not defined workable responses to the increasing presentation of dementia among people with [intellectual disability]” (p. 280) [6]. The Edinburgh Working Group further proposed that “prevailing practices and policies of service provision need to be examined with an eye to adopting universally applicable guidelines which promote continued community care and support of people with [intellectual disability] who are affected by dementia” (p. 280).

2. Guidelines

The WHO has noted that “care needs, which include identification, diagnosis and symptom management as well as long-term support, often challenge the skills and capacity of the workforce and services ... [and] to improve the quality of life of people with dementia and their caregivers, it is essential that the care provided by health and social care services is coordinated and integrated and can be adapted to the changes that occur throughout the course of the disease” (p. 50) [1]. This can be affected by national guidelines, but that such “national guidelines should also be flexible enough to accommodate differences in regions and in cultural groups and, where required, should be translated into different languages and dialects” (p. 62) [1]. The US Institute of Medicine (IOM) defines clinical practice guidelines as those “statements that include recommendations intended to optimize patient care that are informed by a systematic review of evidence and an assessment of the benefits and harms of alternative care options” [7]. The WHO proposed that *clinical practice guidelines* should describe the benefits of early and accurate diagnosis; identify behavioral and psychological symptoms of dementia; cover medical management of dementia, including diagnosis, assessment, and management of symptoms; identify the process of referral for specialist assessment and

diagnosis; recommend nonpharmacologic interventions; and provide for caregiver assessment of support needs. The WHO also proposed that *long-term care guidelines* should provide guidance for social engagement and recreational activities, cognitive stimulation, physiological support, safe and stimulating environments, recognition of behavioral and psychological change and its potential causes, management of behavioral and psychological symptoms, palliative support, and psychosocial support for carers [1].

The first such attempt at guidelines for adults with intellectual disability affected by dementia was undertaken by an international working group in the early 1990s [8]. Currently, a number of country-focused documents serve as guidelines for assessing and caring for people with ID and dementia, but none rise to the level of the criterion of the IOM [9–12]. They do, however, fulfill the criterion of being experiential and consensus documents and address the *initial* WHO content goal for guidelines, that is, they provide guidance for screening, assessment, and diagnosis—crucial areas as there is significant discordance between screening and assessment practices with persons in the general population affected by dementia and for diagnostic practices needed to be used with adults with ID who manifest varied premorbid intellectual skills and capabilities. Typically defined are the types of measures that can be used, the manner of employment, and the interpretation of the outcomes. These guidelines share various commonalities and can aid the general practitioner and others with gaining a level of comfort during the initial and subsequent examinations and having some familiarity with referral sources.

Others fulfill to varying degree the *secondary* WHO goal—as they focus on social care practices that can aid practitioners with referral options for alternatives for community supports and residential care, such as in the UK [9] and in the United States [13]. Typically, guidance is offered on individual care practices as well as congregate settings and aid for family caregivers. Most of these guidelines offer commonalities in their guidance and recommendations within the context of national or regional resources and can be useful for the general practitioner and others in making appropriate referrals.

3. Commentary

Guidelines have a useful function. They can raise practitioner awareness, define assessment and diagnostic processes, source specialists, promote systematic screening, identify foundational supports for family caregiving, stimulate preventative initiatives and systematic early detection, promote health and wellness (via nutrition and exercise), and promote public health planning and the development of community long-term care options. Although extant national guidelines do not rise to the level of the IOM criteria, they currently provide the best sources based on consensus among practitioners and researchers, and do at times have an empirical basis, particularly when providing guidance for screening, assessment, and diagnosis, as well as the use

of pharmacologic applications. Where divergence occurs, it is generally related to variations in national social care practices and options; however, even here, there is more consensus than divergence. Yet, although these individual guideline efforts exist, little effort has been undertaken to arrive at a consensus with respect to universal practice and application—such that would satisfy the call from the WHO.

Thus, we would argue that there is a medical and social benefit for a set of unified and comprehensive guidelines related to ID and dementia that transcend national interests and practices and provide consensus on internationally accepted practice—emanating from an international organization, such as the WHO or Alzheimer's Disease International. Where this is not practical or possible, guidelines developed and representative of national multi-interest groups and associations are the next best course (such as was done in the United States by the National Task Group on Intellectual Disabilities and Dementia Practices—see www.aadmd.org/ntg). Providing information for practitioners, providers, families, and persons affected, guidelines can help standardize the approaches to the variety of efforts being undertaken to detect dementia early in people with ID, help stimulate a research agenda, and guide practitioners with respect to courses of treatment and surveillance of comorbid conditions, as well as guide the social care system in engaging in best practices with respect to community supports and providing quality care for persons with dementia regardless of stage or degree of impact.

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