Practice guidelines for the clinical assessment and care management of Alzheimer's disease and other dementias among adults with intellectual disability*

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Abstract

The AAMR/IASSID practice guidelines, developed by an international workgroup, provide guidance for stage-related care management of Alzheimer's disease, and suggestions for the training and education of carers, peers, clinicians and programme staff. The guidelines suggest a three-step intervention activity process, that includes: (1) recognizing changes; (2) conducting assessments and evaluations; and (3) instituting medical and care management. They also provide guidance for public policies that reflect a commitment for aggressive care of people with Alzheimer's disease and intellectual disability, and avoidance of institutionalization solely because of a diagnosis of dementia.

Introduction

Although information exists on the natural history of Alzheimer's disease among adults with intellectual disability (Dalton & Crapper-MacLachlan 1986; Oliver & Holland 1986; Berg \textit{et al.}, 1993; Harper 1993; Wisniewski \& Merz 1995; Wisniewski \textit{et al.} 1994) and on the means of conducting diagnostic assessments (Alyward \textit{et al.} 1995), information on care practices has been quite limited (Marler \& Cunningham 1994; The Arc 1995,a,b). Because of the nature of the disease, clinicians concur that, when there is confirmation, it is important to begin to modify certain care practices and supports to accommodate the expected changes in behaviour and capabilities (Newroth \& Newroth 1980; Kultgen \& Holtz 1992; Holland \textit{et al.} 1993; Noelker \& Somple 1993).

However, no standards or practice guidelines have been available to help such modifications (Gambert \textit{et al.} 1988; Carlsen \textit{et al.} 1994; Chicoine \textit{et al.} 1994).

To help define such a set of universally applicable practice guidelines, an international colloquium on Alzheimer's disease and intellectual disability was convened in Minneapolis, Minnesota, USA, in 1994 (Janicki 1994; Deb \& Janicki 1995). The colloquium participants, drawn from experts from North America and Europe, noted that the occurrence of Alzheimer's dementia will have a profound impact on many families and social agencies with the growing emphasis on providing personalized supports to adults with intellectual disability in their

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communities and the increasing life-span of people with intellectual disability. Thus, these practice guidelines were developed to provide guidance to professionals and families on the clinical assessment and care management of adults with intellectual disability and possible or probable Alzheimer’s disease.1 What follows is an abridged version of the AAMR/IASSID practice guidelines (for the complete version and fuller explanation of the guidelines development, see Janicki 1995).2 These guidelines can equally apply to adults with intellectual disability with other diagnosed dementias whose course is irreversible, slow and progressive.

**Practice guidelines for care management of Alzheimer’s disease and intellectual disability**

The AAMR/IASSID practice guidelines are based upon the following assumptions: (1) each person’s own needs must be the basis for how care is provided; (2) while some age-associated changes are normal (e.g. changes in stamina and sensory abilities), gross mental deterioration is not; (3) people with Down’s syndrome are at greater risk of Alzheimer’s disease; (4) some behavioral changes may look like Alzheimer’s dementia, but may be the result of other causes and may be reversible; (5) the differential diagnostic process should be essentially the same as that used in the general population, except for modifications made to accommodate differing cognitive skills; and (6) the individual’s own abilities and level of functioning at any point in time (i.e. his or her baseline of performance) should be the basis for evaluating subsequent changes. The guidelines suggest intervention activities in three steps: (1) recognizing changes; (2) conducting assessments and evaluations; and (3) instituting medical and care management. These guidelines provide suggestions rather than prescriptions, and are addressed to various levels of carers and diverse disciplines.

**Step 1: Understanding changes in normal ageing, being aware of risk factors and recognizing changes indicating the onset of dementia**

Normative ageing results in certain sensory, physical, psychological and behavioural changes. To understand pathological changes, it is important to know the differences between these normative changes, and changes that result from disease or other pathological processes. Furthermore, although the staging of dementia symptoms does not appear to differ among people with intellectual disability in general, the timing and manner that symptoms may be expressed can vary widely from individual to individual. These symptoms may also appear differently among adults with Down’s syndrome than among adults with other etiologies of intellectual disability.

Adults with intellectual disability who are at risk of Alzheimer’s disease include those over 50 years of age, those with Down’s syndrome over 40 years of age, or those who are from families with a history of Alzheimer’s disease. The presence of any of these factors does not necessarily mean that Alzheimer’s dementia (or another form of dementia) will occur. However, the presence of one or more of these risk factors may indicate an increased risk of an adult with intellectual disability manifesting dementia.

When these risk factors are present, periodic screenings are particularly helpful for identifying potential changes in behaviour that may suggest pathological ageing. Changes that may be early indicators include: unexpected changes in routine behaviours; a decrement in functional abilities, such as cooking, dressing or washing; memory losses or difficulty in learning new activities; changes in attitude or demeanor; a loss of job or social skills; withdrawal from pleasurable activities; night-time awakenings and other altered time difficulties; increases or decreases in rigid behavioural patterns; and the onset of seizures. Because observable changes in behaviour may be the result of causes other than dementia (e.g. depression, sensory impairments and hypo/hyperthyroidism), and may be treatable and reversible, referral for a diagnostic
workup should be made as soon as possible after observing any of the signs noted above.

A periodically applied screening instrument should be used to establish both a behavioural baseline and to obtain longitudinal measures that indicate change. A baseline screening should be undertaken that includes cognitive, health and functional assessments beginning at 40 years of age in individuals at increased risk for premature ageing, such as people with Down’s syndrome, and beginning at 50 years of age in others. The individual should also receive periodic cognitive, health and functional assessments. These on-going assessments could reveal significant changes in function. Where periodic screenings are not practical or possible, an alternative means of assessing change should be used. One means is to ask the adult to keep a ‘life-history’ diary in which are noted significant events, abilities and documentation of capabilities. Such baseline measures or life-histories can help others understand the adult’s normative behaviour, and thus, any changes that suggest pathological ageing will be highlighted.

Step 2: Conducting assessments and evaluations

When there is suspicion of dementia, referral for a thorough evaluation should be done to assure a proper differential diagnosis. Thus, the second step includes: (1) gathering information on behaviour to further confirm noticed changes, preferably from multiple informants such as staff or carers; (2) continuing to monitor behaviour/functioning for presentation to clinicians; and (3) making a referral for a diagnostic workup for a differential diagnosis. To make a distinction between possible and probable diagnosis of Alzheimer’s disease (McKhann et al. 1984), it is necessary to observe a well-documented progression of symptoms substantiated by appropriate clinical test results. Because periodic observation of behaviour is a critical feature of the diagnostic evaluation among adults with intellectual disability, obtaining a confident diagnosis requires repeated evaluations.3

Resources for general diagnostic evaluations may include geriatric assessment clinics, memory assessment and memory disorder clinics, Alzheimer’s disease assistance centers, Alzheimer’s disease centers, specialist or geriatric health care teams, general practitioners, neuropsychologists, neurologists, geriatric psychiatrists, and other physicians. Other, more specialized, resources may include special clinics of local intellectual disability, mental health or psychiatric, ageing or senior services, agencies, and university programmes in intellectual or developmental disabilities.

Step 3: Instituting medical and care management

The third step involves two interwoven paths: medical management and care management. Contact between these two paths should be routine and ongoing, depending on the individual’s needs.

The medical management path calls for systematic treatment of all treatable medical conditions, such as hearing disabilities, seizure disorders or cataracts. Treatment of these conditions should be as thorough as it is in the general population. Co-morbid mental disorders (such as depression) should be treated aggressively. In particular, the treatment must be tailored to a clearly established diagnosis, rather than to vague behavioural symptoms, such as aggression. The use of psychoactive medications for behavioural control should be limited to acute situations, and should be replaced, when possible by appropriate behavioural, cognitive and environmental interventions.

Frequent review of all medications is necessary, with the goal of using the fewest number and lowest possible doses of effective medications. Continued monitoring of medical, psychiatric and cognitive changes must occur, as conditions tend not to be static but evolve with time. Pharmacological therapies must consider carefully the increased vulnerability of the central nervous system to further cognitive impairments. Particularly likely to cause impairment is the use of multiple medications (polypharmacy).

With progression of the disease, medical management takes on a more intensive course, often inverse to the time spent in care management. When there is an overwhelming loss of personal care

3See Ajeward et al. (1993) for a detailed description of diagnostic criteria, evaluation processes and suggested instrumentation. See also Zigman et al. (1995) for suggested data to be collected during evaluations and information retrieval.
skills and mobility during the last stage, the person may no longer be able to walk, sit up, chew and swallow food, or control bowel or bladder. Added to these losses of function and general unresponsiveness, the person may experience the onset of seizures and is at greater risk of infection. Given the total loss of body functions, practitioners must put more emphasis on primary nursing care and medical management to deter infections and defer death for as long as possible.

The care management path calls for documenting and carrying out a treatment strategy appropriate to each stage of the disease. General principles of care management include: (1) helping the person preserve and maximize function; (2) using interventions and supports that are appropriate to the stage of the disease; and (3) conducting care planning that is multidisciplinary and involves information from multiple sources. Care management involves structural activities such as making referrals for appropriate services, making environmental modifications, changing the general plan of care according to identified sustainable abilities of the individual, and determining whether to use ageing- or Alzheimer's-disease-related services in addition to or instead of intellectual disability services. Care management also involves individualized applications of clinical strategies to address problem management and carer concerns. Written documentation is crucial in this step as it helps identify changes and personal care needs.

Once the suspicion of Alzheimer's disease has been clinically confirmed, the person's family or other carers need to be made aware that what may have been comfortable and familiar for the individual in the past may become unrecognized and result in unpredictable behaviour. Changes may need to be made in daily routine and environment so that the adult can feel safe and secure in his or her environment. Carers should be encouraged to promote this feeling of safety, because, although the adult may be mobile, his or her judgement may be decreasing, and he or she may be at risk of falling or other injuries. Under these circumstances, some of the person's responsibilities may need to be modified or curtailed.

Family, friends and companions (including providers and staff) are integral to care management. They should be used as genuine supports and encouraged to gain a better understanding of the nature and course of the disease. A balance should be maintained between providing supports that compensate for the loss of skills and encouraging the individual to perform activities that may preserve function. As the disease progresses, treatment practices need to be modified to meet the changing needs of the individual with probable Alzheimer's disease. These modifications need to accommodate diminishing self-care, communication, and orientation skills.

Attention should be paid to addressing eating, balance and mobility difficulties, and problems with continence and wandering. Simplifying the environment, establishing a regular routine, and applying common-sense oversight and direction will help address these changes. It may be necessary to reduce the number of alternatives in the person's daily life because making choices can be confusing and frustrating for someone with dementia. While reducing the extent of individual alternatives to minimize confusion, the carer should simultaneously offer a variety of broad experiences drawn from programmes and amenities available within the community. The person should not be isolated, but should be encouraged to continue membership for as long as possible in his or her community. Supports, services, and care management strategies should be designed to accommodate differing needs at the various stages of the disease, including the early, mid- and late stage.

Early-stage practices

As the onset of symptoms often appears very gradually, changes in supports and services may be minimal at first. There may be some small memory loss, particularly of recent events. Adults may have trouble in finding the right words to use during casual conversations. Work performance may begin to deteriorate and changes in behaviour may start to become obvious. Adults may also experience some altered time concepts, loss of familiarity with routine activities, or loss of interest in favoured hobbies, events or activities. There may be periods of lessened alertness and slowing of movement.

Early signs and symptoms of Alzheimer's dementia do not mean that a change of familiar
programme or residence is necessary or even desirable. Maintaining routine and familiarity with the environment can help compensate for any changes or disorientation in the adult's behavior. To the extent possible, the adult should be allowed to 'age in place' with dignity and respect. The adult's programme or home environment should be adapted for safety, ease of access, and ultimately, to maintain or slow down loss of function. Supervision or personal assistance levels should be modified to accommodate changing abilities.

Early stage care practices often include providing supports that buttress the person's ability to enjoy normal activities by: adapting or simplifying the activities to the person's changing cognitive abilities; providing structure and support for performing daily routines; using explicit, short directions or instructions, cues, and verbal prompts; maintaining favourite activities; implementing approaches that match the changing abilities of the individual; and engaging the individual in activities involving other members of the household or programme. Adults who experience changes related to dementia should be involved in activities and exercises that can support their positive sense of involvement, accomplishment and well-being, and specifically aid in the preservation of muscle tone and strength. The practice goal is to optimize the person's sense of success in everyday activities, encourage positive self-esteem, and maintain autonomy and good physical and emotional health for as long as possible.

Mid-stage practices

During this stage, the behavioural changes noted during the early stage become more obvious. Distinct losses of language abilities are frequently the most obvious sign that the individual is progressing to this stage. For example, adults affected may have difficulty naming objects or with maintaining a logical conversation. They may also have difficulty understanding directions or instructions. They often become disoriented as to time, place and person. Memory losses and confusion often lead to frustration. Thus, carers need to acknowledge with enthusiasm what the individual is saying and doing, and to do so using the adult's perceptual field as the point of reference.

There may also be the beginning of a loss of self-care skills and continence. Significant changes in personality and social behaviour begin to appear. These changes are often associated with paranoia and delusions. Late-onset seizures may become evident for the first time. Thus, additional supports such as respite, personal care assistance, physical alterations to the home and more frequent health monitoring at this stage may help continue to maintain the individual in his or her residence and moderate some of the behavioural changes.

Questions may also arise regarding self-determination, advanced directives and guardianships (for a fuller explanation, see NYSOMRDD 1994). Where legally permissible, a health care proxy should be executed authorizing a named individual to make health care decisions after the adult becomes too incapacitated to make his or her own. The proxy should cover artificial hydration, nutrition, extraordinary medical procedures and resuscitation. When a guardian has not been appointed, the adult or carers want to start guardianship, using whatever legal processes exist in the jurisdiction.

Overall, the following practices are useful in mid-stage of the disease: preservation of function; maintenance of physical and dental health; adequate nutrition; protection and maintenance of safety; aid with self-care; participation in stimulating activities; strategies to reduce agitation; and a periodic review of physical function and health for ongoing planning of appropriate interventions. With increasing disorientation, forgetfulness and sometimes personal agitation, greater care should be given to the design of the person's routine, activities and safety. Since wandering, disorientation and agitation, together with a loss of orientation to visual cues, may occur, the person's negotiation of his or her environment is enhanced by using special markings, colors and textures. Continence can be maintained by monitoring fluid intake, timing of voiding and ensuring that toileting facilities are carefully marked.

Carers should ensure that adult is maintained on a balanced diet and receives adequate nutrition by using techniques that encourage safe eating, including using appropriate food consistency and

*See Olsen et al. (1993) for ideas on environmental modifications.

portions, allowing adequate time for eating, and taking advantage of times when the person is not fatigued. Maintaining flexibility in nutrition management, such as simplifying meal routines, avoiding excess stimuli during meal time, providing finger foods and more frequent smaller meals, and supplementing dietary needs by vitamins or healthy snacks can help maintain good nutrition and safe eating as the disease progresses.

Some families or other carers may benefit from respite and information on care management techniques. Persons helping carers cope should be aware of carer stresses or problems associated with lack of relief for caring: coping with their own health difficulties and caring for others in the household. Carers in these situations may also benefit from counseling and future planning, particularly as the adult continues to lose function.

Late-stage practices

Late or end-stage needs require special attention and sensitivity. During this stage, adults experience substantial dysfunction. Basic skills such as eating or drinking are lost. Because of eating problems, activity level and changes in metabolism, many adults may experience a substantial loss of body weight. They may eventually lose their ability to maintain balance and walk. Their long- and short-term memories are typically lost, as is their ability to recognize other people and their environment. At the very late stage, people affected require 24-h care and supervision. They often become bedridden and inactive. Because they are bedridden, they are at increased risk of any infection, especially pneumonia, and consequently, are far more likely to die. Special care must be exercised to prevent dehydration, choking or aspiration pneumonia, and skin pressure sores. Preventive care may curb these conditions, and thus, support the maintenance of comfort.

In this stage, adults also lose their ability to care for themselves, and bowel and bladder incontinence becomes a problem. Previously simple activities like eating, washing and grooming will require extensive personal care attention. Adults will require constant direction and supervision, and will generally not be able to be left alone. When adults are still able to walk, wandering presents a significant risk to safety and health. If excessive wandering cannot be prevented, environmental modifications, such as special pathways, or targeted supervision needs to be designed into the programme. With advancing decline, adults may be lacking any affect and have a complete lack of awareness of their surroundings. Although most verbal abilities may be lost, some use of words or phrases may be retained. When this occurs, carers should consider methods for retaining these abilities.

Late-stage interventions should also attend to the problems of carers burdened by the strain of caregiving. At this stage, more effort should be given to supporting the family or other carers directly involved caring for the individual. Special consideration should be given to helping staff and carers handle bereavement and deal with the stresses inherent in providing terminal care. Use can be made of clergy or other spiritual supports, and also hospices.

Education, training and care practice policies

To promote early recognition and referrals, clinicians, workers, carers and peers should receive training in normal ageing processes and indicators of change signalling a dementing process. Both the ageing and disability community should be targeted for such training. To promote appropriate care, training should be provided in care management techniques and the specific approaches for coping with functional limitations and death. The organizing principle underlying this training is the individuality of the adult with dementia, and a perspective that promotes personal dignity, autonomy and personal welfare.

Family oriented materials should be produced and made available locally to help carers to obtain information and assistance. Training should be provided to family carers to enable them to more effectively maintain their relative’s functioning and to know how to seek needed services. Information about programme supports, such as day services, respite and in-home services, should be made available. As appropriate, families should be connected to support organizations for Alzheimer’s disease or for intellectual or developmental
disabilities, and others providing opportunities for support. Diagnostic and practice information should be made available throughout the professional community (e.g. see McLennan et al. 1993) and to family carers (e.g. see Marler & Cunningham 1994; The Arc 1995a, b) in a variety of media. Physicians and other workers at acute, managed and long-term care facilities should be familiar with how the dementing process is manifested in adults with intellectual disability and how to carry out effective evaluations. This training is important at both the pre-service and continuing education levels.

Workshops and training courses should provide information on Alzheimer’s disease diagnostic and treatment practices with particular application to people with intellectual disability. These workshops and courses should, at minimum, contain information on normal ageing, Alzheimer’s disease, recognition of early signs of dementia, methods for conducting periodic assessments and evaluations, available services, supporting carers, general care management, and effective practices for early, mid- and late-stage interventions.

Agency and public policies must recognize that contemporary and future research on genetic testing may provide reliable advance warning of susceptibility to or risk of Alzheimer’s disease. Legislation or policy that prohibits discrimination based on findings of such test results should be sought and followed. Agency care practice policies must reflect a commitment for aggressive care of persons with Alzheimer’s disease and intellectual disability and avoidance of institutionalization based solely upon a diagnosis of dementia. Training on alternate approaches of care should be emphasized so that care practices are consistent with these policies.

AAMR-IASSID Practice Guidelines

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