DIAGNOSIS AND MANAGEMENT OF DEMENTIA AMONG ADULTS WITH INTELLECTUAL DISABILITY

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Preparing for the Future: Alzheimer’s Disease and Related Dementias
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WHO report on dementia

- Prevalence and incidence projections indicate that the number of people with dementia will continue to grow, particularly among the oldest old.

- The total number of people with dementia worldwide in 2010 is estimated at 35.6 million and is projected to nearly double every 20 years, to 65.7 million in 2030 and 115.4 million in 2050.

- Primary prevention should focus countering risk factors for vascular disease, including diabetes, midlife hypertension, midlife obesity, smoking, and physical inactivity.

Intellectual disabilities

Care pathways for populations with specific needs
- ‘Some groups have additional needs arising from having dementia or being a caregiver of a person with dementia. Examples of specific or minority groups include... people with intellectual disabilities...’
- ‘Some of the barriers to access include a lack of understanding or recognition of the dementia in their population group, language or cultural barriers, and a lack of appropriate information resources and services.’

Intellectual disability
- ‘People with Down syndrome are at a significant risk of developing Alzheimer’s disease. Studies suggest that 50–70% will be affected by dementia after the age of 60 years.
  - The onset of dementia in people with Down syndrome is likely to be younger than the sporadic form of dementia that generally affects older people.’
- ‘In the USA, a national task force of experts on intellectual disabilities and Alzheimer’s disease has developed a comprehensive report with policy and practice recommendations on detection, care and support for this population.*
  - The aim of the report is to enable adults with intellectual disabilities who are affected by dementia to remain living in the community with quality support.’

2014 Update to the National Plan to Address Alzheimer’s Disease

Activities – having implications for intellectual disabilities – directed toward…

- Developing targets, strategies, and milestones for dementia-capable primary and specialty care workforce
- Continuing work on measuring high-quality dementia care and identifying meaningful outcomes
- Exploring models of family support for people with intellectual disabilities as they age
- Developing a compendia of state policies on residential care and adult day health
- Enhancing public awareness and engagement (national education and outreach initiative)
- Producing a chartbook on characteristics of people with AD and their caregivers
Demographics are trending upwards for a larger older population.

Alzheimer’s disease currently affects 5.2 million Americans (and 9 million by 2050)\(^1\)

In more than 90% of adults with Alzheimer’s disease, symptoms generally appear after age 60, and the incidence of the disease increases with age\(^1\)

The numbers of older adults with ID are growing as is the number affected by dementia

\(^1\) source: Alzheimer’s Association
Needs...

- Need for ‘best practice’ models in community based housing
- Need for supportive services when in family home
- Need for practice guidelines for medical and health care settings
- Need for screening, assessment, and diagnostic resources
- Need for more information on population and effects of dementia on long term care
Diverse older population – by urgency of needs

Well-elderly – main needs are social and generic health … deferred care needs

Marginal needs – Some aging related issues and changing demands for care/supports … aging-in to elder care

Most at need – age and disability associated impairments … immediate and intense care needs… adults with dementia
Dementia and Intellectual Disability
Dementia & ID: the questions

- What is dementia? How does it differ from lifelong intellectual disability?
- How does it impact people with intellectual disabilities? Is the impact different?
- Will all persons with ID be affected in old age? Why and who will?
- How can we respond? What services are needed? Who can help us?
ID vs. Dementia

- Intellectual disability involves undeveloped or underdeveloped mental or intellectual skills and abilities.

- Dementia is a widespread loss of mental or intellectual skills and abilities. Dementia-related losses occur in memory, language skills, orientation, ADLs [activities of daily living], and changes evident in personality and global functioning.

*Key:* behavioral presentation may be similar, but it’s the loss from previous level of function that differentiates the two.
Dementia and ID

Onset

Progression

Duration
Differences in ID?

Early onset?
- Among some etiologies – e.g., Down syndrome, head injury

Symptom presentation?
- Mostly the same, some variations due to etiology – e.g., Down syndrome… early personality losses

Assessment?
- Standard measures not useful – comparisons by individual over time

Services?
- More dementia care options as lifelong supports generally available
When Does Onset Occur?

Onset is speculative . . .
- the best we can do is identify that point when significant change or impairment has become noticeable
- in some people noticeable changes have been proceeded by MCI – “mild cognitive impairment”
- early 50s for DS – late 60s for ID

Note difference between DS (yellow) and other ID (green)
Onset is speculative . . .

- the best we can do is identify that point when significant change or impairment has become noticeable.
- in some people noticeable changes have been proceeded by MCI – “mild cognitive impairment”
- early 50s for DS – late 60s for ID
Progressive cognitive deterioration due to Alzheimer’s disease

Stages of Alzheimer’s Disease

**EARLY or MILD STAGE**
2 to 4 years or longer

**MID- or MODERATE STAGE**
2 to 10 years

**LATE or SEVERE STAGE**
1 to 3 years of longer

Critical factors

- Degree of retention of function
- Expected trajectory of progressive dysfunction
- Duration (remaining life years)
- Type of dementia
- Health status
- Environmental accommodations

Varying trajectories have implications for continual assessment and adaptations to care management

Source: Figure 1 from Wilkosz et al., (2009). Trajectories of cognitive decline in Alzheimer’s disease. *International Psychogeriatrics*, 28,1-10
## Dementia & ID Findings

### Rate of Occurrence

<table>
<thead>
<tr>
<th>Source</th>
<th>Description</th>
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- Age-cohort percent for adults with intellectual disability (ID) is same as in general population (~5-6% over 60)
- Much higher prevalence (60% >age 60) and neuropathology indicative of AD in most adults with Down syndrome (DS)

### Dementia Type

- Type of dementia differs between DS and other types of ID
- Generally dementia of the Alzheimer’s type is prevalent in DS
- Similar range of dementias found in ID as in other people
- Problem of ‘diagnostic overshadowing’ in DS

### Onset and Duration

- Average onset age in early 50s for DS – late 60s for others
- Most DAT diagnosed <3 years of “onset” in adults with DS

### Behavioral Changes

- More evident change in personality in DS
- More initial memory loss in other ID

### Neurological Signs

- Late onset seizures found in 12%-84% of adults w/DS

### Prognosis

- Aggressive AD can lead to death <2 years of onset in DS
- 2-7 years mean duration in DS
- Same duration expected among other ID as in other people
What’s important to know

- The difference between normal aging changes and pathological aging changes
- Early signs of functional change associated with dementia
- Types of dementia and their main characteristics, what will be the behavioral/functional changes, and their duration
- When is it best to refer for assessment and to whom
- What options exist for early dementia-related supports
- What options exist (or need to be put in place) for long-term dementia capable care/supports
Dementia Detection and Adaptation Model

Suspensions are raised or routine screening detects changes
Assessment request is made
Assessment validates suspicions and changes
Reassessments indicate continued decline and changes
Diagnostic review is requested
Differential diagnosis is made

Examples...
Short term memory loss
Confusion
Decline in abilities
Challenging behaviors
Personality changes
Wandering
Falling

Assessment by psychiatrist, neurologist, psychologist

Program/services adaptations

Dx: Possible or probable dementia

Dx: Not dementia

Changes at residence

Diminishing function
Stage related concerns

Dietary changes – hydration, nutrition, weight
Changes in staff – staff paired to dementia stages
Adaptations of living environment – sounds, lighting, way-finding, safety
Adaptations to routines and activities – lower stimulation, but engaging activities
Specialized attention to ADLs, health, mental health, incontinence, memory cues
What to do when dementia is suspected?

- Apply a detection/screening instrument to look for signs
  - if comes up positive
- Refer for assessment...
  - To whom?
    - Agency MD, local MD, psychologist, nurse, other person who may do formal assessment to validate suspicions
  - If outcome comes up positive
- Refer for diagnostic work-up
  - Ask for confirmatory information and determination that it is dementia (*by exclusion of other causes*)
Ascertaining dementia

- Start with an administrative screen (e.g., NTG-EDSD)
  - Capture visuals on functioning (preferably ‘personal best’)
    - digital recording of behavior
  - Observe if screen provides ‘hits’ on ‘warning signs’

- Refer for clinical assessment
  - Clinicians reapply ID-specific measures to look for longitudinal patterns
  - If evidence points to dementia-like symptoms, refers for full diagnostic evaluation (for differential dx)

- Obtain diagnosis and track function
Benefits of differentiating types of dementia as part of the dx process

- Diagnostic precision
- Potential medication treatment variations
- Developing expectations of residual life years
- Setting up care management plans on expected behavioral presentations and progression
- Communication and interaction variations
- Projecting expectations for change in care needs
Challenges to ID diagnosis and care

- Individuals with ID may not be able to report signs and symptoms
- Subtle changes may not be observed
- Commonly used dementia assessment tools are not relevant for people with ID
- Difficulty of measuring change from previous level of functioning
- Challenges with assessing change in adults with severe and profound ID
- Conditions associated with ID maybe mistaken for symptoms of dementia
- Diagnostic overshadowing
- Aging parents and siblings (informants or carers)
- Dearth of research and training on differential Dx
Screening

Assessment

Diagnosis

Administrative [NTG-EDSD]
Clinical (DSDS, DLD, AADS)

Dementia Assessment Scales
Neuro-psych evals
CT - MRI

Possible
Probable
Definitive
If you are concerned as to whether it is dementia...

- Keep track of the behavior
  - Use a digital recording device
  - Use a screen/early detection instrument
  - Seek advice from another clinician
  - Gather as much information as you can
    - Speak with other program staff
    - Speak with housemates, etc.
    - Speak with family
  - Review record for previous status
    - History
    - Mention of decline
Need for a early detection/screening tool

- Clinicians report individuals often have problems well in advance of significant signs of dementia

Why screening?
- Screening tools capture information about early changes in behavior and function
- Caregivers can be good observers and reporters of relevant signs and symptoms of changes
- Screening tools provide a common format for sharing information

- Cognitive and functional status are not usually included in periodic health exam visits
  - Relevance to ACA cognitive assessment as part of the annual wellness visit
Early & Periodic Screening

- Applying screening/detection and assessment measures
  - Controversy as to whether this is intrusive
  - Balance privacy vs. risk
- Benefits
  - Pick up on early warning signs
  - Helpful with early differential dx
  - Associate with data from biomarkers
- Easy ways to collect quick impressionistic data
- Protocol for recording a set of signal performance tasks and behaviors linked to potential early expression of dementia
- Can be used as part of the individual’s permanent record or program plan to periodically track changes in behaviors that might indicate cognitive and motor changes associated with dementia

Available at http://www.uic.edu/orgs/rrtcamr/recordingbehav.htm
Early detection/screening

‘NTG-Early Detection Screen for Dementia’ (NTG-EDSD)

- Usable by support staff and caregivers to note presence of key behaviors associated with dementia
- Picks up on health status, ADLs, behavior and function, memory, self-reported problems
- Available in several languages

Use: to provide information to physician or diagnostician on function and begin the conversation leading to possible assessment/diagnosis

http://aadmd.org/ntg/screening
‘NTG-Early Detection Screen for Dementia’ (NTG-EDSD)

Usable by support staff and caregivers to note presence of key behaviors associated with dementia

✓ Picks up on health status, ADLs, behavior and function, memory, self-reported problems

✓ Available in several languages

Pages ①②: Basic information

Pages ③④: Information about function and indicators of problem areas associated with dementia

Page ⑤: Coincident conditions

Page ⑥: Medications & Comments
Assessment

- **Function**
  - To examine adults due to suspicion of cognitive or functional decline that may result from Alzheimer’s or other conditions

- **Process**
  - Collect informant information (family, staff, friends, etc.)
  - Apply detailed measures assessing function, and physiological and neurological factors
  - Conduct collateral examinations (neurological, CT/MRI)

Diagnostics...

- Focused medical and family history
- Physical exam and laboratory tests
  - Functional status exam
  - Mental status exam
- Assessment for delirium/depression
- Brain scans (CT or MRI)
- Neuropsychological tests
Absence of specialized training among clinicians

Poor knowledge of assessment protocols to assess dementia

Dearth of assessment services

Lack of familiarity with identifying dementia in adults with ID among primary care medical practitioners

Confusion by symptoms and misdiagnosis

Reversible/treatable causes of dementia-like behaviors

- Adverse drug reaction
- Depression
- Metabolic changes
- Nutritional deficiencies
- Head injuries
Program options
Dementia and ID

Foundations

Supports

Care management
What is optimal dementia care?

Foundation –
- Settings need to be ‘dementia friendly’ and ‘dementia capable’
- Avoiding institutionalization or re-institutionalization in large congregate settings irrespective of auspice (aging, disability, health)

Structure –
- Individualized, personalized care
- Comfortable settings, offering long term commitment to care, with family or small number of co-residents, capable staff, educated caregivers, adapted environment
- Protection from abuse, harm and neglect, safety
Community Supportive Care

- Dementia lessens an individual’s ability to be left alone – thus eventual ‘living without supervision’ is not an option.

- What are the supports needed?
  - Help for carers and the person
  - Advanced planning for alternative care
  - Diagnostic and intervention assistance
  - Support groups for carers (family or staff)
  - Dementia capable community housing (group homes)
  - Respite for carers
  - Health care and social supports
Dementia Care Focus

Environmental supports
- Compensate for cognitive losses and disorientation
- Support safety and mobility
- Offer protection against injury and disease
- Minimize behaviors that disrupt the equilibrium of the home or pose a danger to self or others
- Help maintain dignity and skills

Primary care practices

Program applications
Program Adaptations

Rethinking how daily life and regular activities are approached

Lessening stimulation, while promoting involvement and challenge

Modifications in communication methods

Supporting residual skills
Coordinated dementia-related supports/services

- Adapting housing
- Dementia capable housing

- Surveillance of symptoms; review of co-incident conditions; follow-along

- Joining specially trained clinicians (MD, RN, ψ, other) to aid staff and families

- Respite, home adaptations, support groups, counseling, education
Factors aggravating care decision-making

Change of care focus

Going from making gains to that of maintaining function and dealing with eventual loss and decline

Implications

- Care staff trained in developmental and growth techniques have to be reoriented to think in terms of maintaining capabilities
- Care situation planning has to consider progressive decline and greater inadequacies
- Care environments have to offer structure, comfort and familiarity

- Much higher prevalence of neuropathology indicative of AD in most older adults w/Down syndrome (DS)
- Alzheimer’s type dementia is most prevalent among adults w/DS – with implications for course and duration
- Studies show average onset age in early 50s for DS and late 60s for other ID
- Dementia presents with variable rates of decline
- Duration may range from 1-2 years to 10-20 depending on type and other confounding factors
- If living alone, self-care not a viable possibility with progressing loss of function
- If living at home, long-term care may not always be possible (caregiver burden, home inadequacies, etc.)
- If living in group quarters, staff may not be prepared to provide long-term care
- Communities may not have viable in-community care resources
Prevalent dementia care options and their intent

**Institutional care**
- [long term care facilities, nursing homes, old age homes, dementia special care units]

**Family care**
- [living with family, other relatives, or other family members of carers]

**Neighborhood group homes**
- [generic group homes, specialized group homes]
  - Group homes for persons with ID who age in the homes
  - Group homes for specialized dementia care

**Sheltered dementia care**
- [assisted living, dementia care homes]

**Carer supports**

**Small personalized care settings**
Value of family home care

- Care at home enables continuity with familiar setting and people
- Family commitment to supporting aging and problems associated with decline
- Value enabled if family gets supports for continued home care (respite, home modifications, aides to help primary caregiver, financial assistance)
- Usually provides safety and security
Value of small group home care

- In lieu of care at home, next best option of comfort for families
- Many countries are adopting small group living as alternative to institutional care for persons in general population
- Intuitively and empirically viable – provide personalized care, in-community living, and connections with familiar settings
- Significant research base on structuring quality settings
- In ID field, readily available and adaptable for age-associated community care when dementia threatens independence
- Models exist for transitioning homes as dementia-capable living arrangements
Prevalent models of group home-based dementia care

**Linear adaptations and care**

- **Early**
- **Mid 1**
- **Mid 2**
- **Late**

**Sequential adaptations and care**

- **Early**
- **Mid 1**
- **Mid 2**
- **Late**

### AGING-IN-PLACE
- Single care home and stable stay

### IN-PLACE-PROGRESSION
- Multiple care homes & movement with progression

Mid = mid-level

Source: JANICKI (2010)
Construct of small home-oriented dementia capable care

- **Fixed staffing** – familiarity, mutual comfort, person focus
- **Unregimented care** – personalized attention and schedules, ‘being at home’
- **Familiar surroundings** – easily navigable, fixed personal space, intimate public spaces, adapted fixtures, lessens confusion
- **Small resident size** – familiarity of others, friendships, home feel, high comfort level
- **Care practices** – tailored to individual, attention to needs, easy pick-up on change indicators, group problem solving
Starkey’s Goebel LIGHTHouse Project consists of three specialized homes for 15 people with intellectual disabilities and dementia.

The three 3,700 square foot (343.7 sq m) homes have five bedrooms each, bathrooms, and shared dining and living spaces.

The homes were designed to provide a supportive community living experience for people with disabilities and dementia with specialized staff support until skilled nursing care is required.

Phase I Study tracked 15 residents and 15 controls (who lived in other settings), as well as examined staff and administrative factors over a three-year period.

Study principals: M. Janicki, P. McCallion, L. Force
GH1 = Diana House; GH2 = WOW House; GH3 = Latimer House
Two years later ...

- Evidence of change in function and increasing health problems or less ‘wellness’
- Residents in homes 2 & 3 showed the greatest impact of dementia over the two years
- Higher number of co-morbidities among dementia residents compared to controls
- Staff time spent on caregiving much more than that for ‘the controls’
- Trending toward individual home specialization as to level of care

Phase II: Longitudinal study [2014-2018] of agency experience with the three community based dementia care group homes
Building capacity and understanding....

Resources
Community care

What needs to be considered?
- Where the person is with respect to dementia progression
  - Can he or she stay where they currently live?
  - Should a specialty dementia-care home be considered?
    - If so, what is the most appropriate pairing?
- What are their wishes (or the wishes of the family)?
- What options exist in agencies or in the community?
- Are current services geared up for long term care?
  - Have a dementia-capable residence?
  - Have direct support and clinical staff knowledgeable of dementia and long-term dementia-related care?
Community care

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Social advocacy
National Task Group on Intellectual Disabilities and Dementia Practices

- To define best practices that can be used by agencies in delivering supports and services to adults with intellectual disabilities affected the various dementias
- To identify a workable national a ‘first-instance’ early detection / screening instrument
- To produce educational materials of use to families, people with ID, and providers of services
- To further public policy with respect to dementia as it affects adults with intellectual disabilities

www.aadmd.org/ntg
NTG Activities

- An early detection-screening instrument (NTG-EDSD) & manual
  - Various language versions available
  - Access at www.aadmd.org/ntg

- Practice guidelines
  - Community supports and health practitioner assessment guidelines issued
  - Several others pending
    - Administrative practices, day-to-day care
  - Program standards pending

- Training and education activities
  - Training workshops
  - Meetings with professional groups
  - Information for families (FAQ)

- Linkages
  - US Administration on Community Living
  - NASDDDS, N4A, NACDD & state activities
  - CARF & national program standards
Need for practice guidelines

- Practice guidelines are a formulary – they structure approaches to care – offering guidance on what, where, and when
- Help define how to approach challenges in care, from individual interactions, to program practices, to assessment and interventions
- Stem from consensus and experience as well as evidence-base
- Form basis for regulatory compliance
Guidelines for Community Care and Supports for People with Intellectual Disabilities Affected by Dementia

The guidelines suggest what actions should be undertaken to optimize community-based care and supports.

They follow reflect the progressive nature of prevalent dementias and use a staging model generally accepted for practice among generic dementia services...

... from a pre-diagnosis stage – when early recognition of symptoms associated with cognitive decline are recognized -- through to early, mid, and late stages of dementia

... and characterize the expected changes in behavior and function

Use: to help providers organize and deliver quality care and supports to people with ID affected by dementia

www.aadmd.org/ntg/guidelines
NTG Practice Guidelines for the Evaluation and Management of Dementia in Adults with Intellectual Disabilities

The NTG’s recommended nine-step approach for assessing health and function.

- Taking thorough history, with particular attention to "red flags" that potentially indicate premature dementia such as history of cerebrovascular disease or head injury, sleep disorders, or vitamin B12 deficiency
- Documenting a historical baseline of function from family members of caregivers
- Comparing current functional level with baseline
- Noting dysfunctions that are common with age and also with possible emerging dementia
- Reviewing medications and noting those that could impair cognition
- Obtaining family history, with particular attention to a history of dementia in first-degree relative
- Noting other destabilizing influences in patient's life such as leaving family, death of a loved one, or constant turnover of caregivers, which could trigger mood disorders
- Reviewing the level of patient safety gleaned from social history, living environment, and outside support
- Continually "cross-referencing the information with the criteria for a dementia diagnosis"
Health advocacy guidelines

- These NTG guidelines are designed to help family members or paid staff prepare for and advocate during health visits and enable provider and advocacy organizations to undertake dementia-related health advocacy preparation and assistance.
  - They define how family caregivers recognize and communicate symptoms, as well as find appropriate practitioners familiar with the medical issues presented by aging adults with lifelong disabilities.

- What’s important in health advocacy?
  - Noting changes in behavior and function
  - Being prepared with items to discuss
  - Raising suspicions with a health care practitioner, during routine or ad hoc visits
  - Helping focus the examination and potentially validate that the decline is the result of the onset or progression of dementia.
  - Following-through on post-exam recommendations
Need for standardized training curriculum

- Curriculum models help standardize what and how critical information is conveyed
- Consensus-based content as to what should be included and what would be most helpful to caregivers
- Training enhances competencies in staff and quality care environments
- Subject to measures of knowledge and basis for evaluation of quality of care
New national curriculum currently in development

Curriculum will have several levels

• **Level 3**: Core curriculum for staff employed in programs directly serving adults with ID and dementia
  - extensive content on in-depth knowledge of dementia and dementia capable care
• **Level 2**: Content for staff employed and participating in a continuing education program
  - content on basic knowledge of aging and dementia
• **Level 1**: Content for new hires participating in orientation and in-service training
  - content on basics of aging, touching on awareness of dementia symptoms; limited time
Where are efforts going?

- Improving understanding of dementia
- Alerting to risk and early signs
- Adapting living environments
- Helping with futures planning (health and social care)
- Aiding families who are carers
- Enhancing staff skills – training with respect to dementia
- Quality checks in services
- Providing stage related services
- Getting federal government to invest resources

- Our experience… All begun as workable endeavors, are works in progress, or are on the planning table
Impact....

- Have lives been improved?
- Have agencies been able to deliver more appropriate and worthy services?
- Are families better off?
- Have we set a standard for how people with ID affected by dementia should be seen and treated?
- Has there been a shift in thinking... and in actual program changes?
- Has there been more attention paid to ID issues within the Alzheimer’s communities (researchers, advocates, public policy people, etc.)?
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www.aadmd.org/ntg

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