

Quality Care for People with Intellectual Disability and Advanced Dementia: Guidance on Service Provision

Mary McCarron, Trinity College Dublin, Dublin, Ireland

Philip McCallion, University of Albany, New York, New York

Karen Watchman, University of Stirling, Stirling, Scotland

Matthew P Janicki, University of Illinois at Chicago, Chicago, Illinois

Antonia Coppus, Radboud University, Nijmegen, The Netherlands

Kathy Service, Northampton, Massachusetts

Juan Fortea, Catalan Foundation for Down Syndrome, Barcelona, Spain

Mary Hogan, Eliot, Maine

Evelyn Reilly, Daughters of Charity Disability Support Service, Dublin, Ireland

Sandy Stemp, Reena Foundation, Toronto, Ontario, Canada

and the Advanced Dementia Working Group of the International Summit on Intellectual
Disability and Dementia

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Abstract

Purpose of Report: The International Summit on Intellectual Disability and Dementia (Glasgow, Scotland; October 2016) noted that advanced dementia can be categorized as that stage of dementia progression characterized by significant losses in cognitive and physical function, including a high probability of further deterioration and leading to death. The questions before the Summit were whether there were similarities and differences in expressions of advanced dementia between adults with intellectual disability and adults in the general population.

Findings: The Summit noted challenges in the staging of advanced dementia in people with ID with the criteria in measures designed to stage dementia in the general population heavily weighted on notable impairment in activities of daily living. For many people with an intellectual disability (ID) there is already dependence in these domains generally related to the individuals pre-existing level of intellectual impairment, i.e., totally unrelated to dementia. Hence, the summit agreed that it is imperative that change is measured from the person's prior functioning in combination with clinical impressions of decline and of increasing co-morbidity including particular attention to late onset epilepsy in people with Down syndrome. It was further noted that quality care planning must recognize the greater likelihood of physical symptoms, comorbidities, immobility and neuropathological deterioration.

Summary: The Summit recommended an investment in research to more clearly identify measures for ascertaining advanced dementia, inform practice guidelines to aid clinicians and service providers, and identify additional markers that may help signal decline and progression into advanced dementia among people with various levels of pre-existing intellectual impairment.

Introduction

The 2016 International Summit on Intellectual Disability and Dementia, composed of individuals and representatives of numerous national and international organizations examined several key topics related to adults with intellectual disability (ID) living with dementia. Summit workgroups were charged to examine topic issues related to policy, services, and clinical aspects. One workgroup was charged to examine advanced dementia and how it factors into care practices for people with intellectual disability. Advanced dementia is defined as the later stages of dementia are characterized by memory loss, problems with communication, loss of mobility, eating and weight loss, problems with continence, and unusual behaviour. This effort resulted in several Summit consensus statements, one that offered guidelines for general care in advanced

dementia,[1] and another that spoke specifically to end-of-life care in advanced dementia[2]. The Summit also noted the need for a complementary statement on practical approaches on how the guidelines might be implemented and applied to both quality of life and quality of death for people with an intellectual disability with advanced dementia. This paper addresses this aspect and proffers a framework for quality care for people with advanced dementia.

The Summit recognized that Alzheimer's disease (AD) is the most common cause of dementia and as the disease progresses the brain degenerates and compromises behaviour and function. Among individuals with an intellectual disability, adults with Down syndrome (DS) are at particular risk as they have an added genetic risk loading for Alzheimer's disease. Prevalence rates for adults with DS aged 60 years and older are higher than that reported for the general population at 60-80%,[3-8] versus 3.1% to 7.6 %, reported for the general population,[9-10] Additionally, the onset of AD occurs at a much earlier age with many adults showing symptoms in their late 40s or early 50s [11-16]

Diagnosing dementia in people with an intellectual disability has proven more complex than in the general population due to varying levels of pre-existing intellectual impairment and increased risk of other health conditions which often mimic dementia (such as, hypothyroidism, sensory impairments, sleep apnoea, B₁₂ and folate deficiency, and depression). There are many challenges in the in the assessment of dementia in people with Down syndrome and with other ID, there is often little knowledge of appropriate screening and assessment instruments and insufficient understanding of how to both measure and interpret changes. Staging of dementia in people with an intellectual disability is also often more challenging because for some decline is often compressed and of shorter duration. The identification of progression to advanced dementia is also more challenging for much the same reasons. Because of these challenges when symptoms of dementia are suspected it may already be too late to engage to engage the person with ID in discussions of their preferences. It is critical therefore that throughout adulthood there be an on-going process of documenting and updating stated preferences and choices.

The utility of general population approaches to defining latter dementia staging that combine clinical impressions with data from instruments, such as the Global Deterioration Scale

(GDS)[17] and the Functional Assessment Staging Scale (FAST)[18] has yet to be established as useful for people with intellectual disability [1-2]. This also means that people with intellectual disability are at risk of either being categorised at a more advanced stage of dementia than is the case, or that the advanced stage of their dementia is poorly understood and is not recognized. Indeed, the utility of these scales as predictors of survival in the general population has yet to be established [19]. Knowing when someone transitions to advanced dementia is important from a care and resource perspective.

[Consensus Statement on Quality Care and Advanced Dementia](#)

A Summit Consensus Statement [1] concluded that decline and staging of dementia in people with ID is best achieved by assessment approaches which incorporate (1) objective comparisons with prior levels of functioning, (2) key informant reports of changes in mood, behaviour and day to day functioning, (3) observations of new late-onset of seizures and (4) signs of increased co-morbidity and frailty that co-exist with advanced dementia. The Summit further envisioned that in a multi-stage framework for quality care, decision-making in the latter stages of decline, must be based on the individual's care preferences, surveillance for signs and symptoms of terminal decline and pending death, and integration of desired care with health related and palliative service collaborations.

[Understanding when dementia has reached a terminal /end stage](#)

Diagnosis and staging are not the only issues of concern. There is often poor understanding, recognition, and preparation on how to address the clinical complications inherent in advanced end-stage dementia. Clinical features of advanced dementia in people with intellectual disability are similar to those of the general population,[20] with the exception that among adults with Down syndrome, new-onset seizures are an additional clinical feature reported in up to 84%,[5,8,21-23]. This compares to 10-22% of new onset seizures reported for the general population [24]. It is increasingly recognized that the clinical picture of end-stage dementia for people with and without intellectual disability includes progressive worsening of memory, confusion and disorientation, respiratory difficulties, recurrent infections, an inability

to verbally communicate, immobility, total support for activities of daily living (such as bathing, washing, dressing), nutritional deficiencies, swallowing difficulties with propensity to aspirate, and ultimately, unresponsiveness and coma [1,2,8,12,20,25,26]. Like the general population, clinical evidence suggests that people with an intellectual disability will often experience multiple recurrent infections (particularly chest and urinary tract infections), worsening of dysphasia and aspiration, and episodes of acute delirium. Meal time difficulties represent a difficult crossroads for families, support staff, and clinicians [27]. Chewing and swallowing problems, agitation and distress, spitting, and aspiration can be traumatic and contribute to stress experienced by the person, the family, and caregivers [28].

The Summit noted that because specialized advanced dementia care is about more than the last six months of life, there is a need for more general guidance on all phases of advanced dementia care and in particular in the focus of care.. It was recommended that in providing supports, care providers distinguish between “futile” and “comfort” care.

‘Futile care’ is defined as care that is no longer able to serve its normal purpose. It may be justified to discontinue such care when there are no physical or spiritual benefits and when such care runs the risk of becoming increasingly burdensome for the individual and serves only to prolong the act of dying [29]. However, it is important to recognize that determining the futility of a specific treatment for a particular person and deciding upon treatment continuation or discontinuation is often challenging. Futility should be determined at an individual level based on the characteristics of each unique situation. Factors to be considered are healthcare professionals’ and individuals’ value systems, goals of care, sociocultural and religious context, and individuals’ emotions and personal characteristics.

Comfort care is “underpinned by a palliative approach which aims to improve the quality of life of the person and their family facing the problems associated with life-threatening illness, through prevention and relief of suffering by treatment of pain and other problems, physical, psychosocial and spiritual”[30].

A Summit consensus statement specifically on end-of-life and advanced dementia recognized that end stage or advanced dementia and related end-of-life decision-making is an

emotional and value laden time with complicated, often long-term, close relationships and bonds with staff and family which can result in distress and disenfranchised grief [2]. Additionally, previous avoidance of end-of-life planning can further complicate decision-making at this critical juncture in care and treatment [27]. These challenges in care provision led the Summit workgroup to both target staging concerns and make recommendations for quality care when the advanced stage of dementia is established. The Summit concluded that for persons with an intellectual disability, and particularly in those adults with Down syndrome, there is limited research evidence on the utility of the FAST/GDS criteria commonly used for the staging dementia,[1] and Mitchell and colleagues have noted that even for the general population such measures are not recommended for estimating survival time of less than 6 months [20]. Further, with compressed staging among adults with DS, the use of normative indicators of transitioning to advanced dementia, often presented in terms of years associated with each stage may be further called into question.

Facilitating Discussions and Thinking Ahead

The Summit participants recommended that discussions of anticipating profound decline and transitioning to global incapacity be a natural extension of the regular Person Centred Planning (PCP) process that occurs in other phases of the person's life [31]. These discussions should address: (a) what and who is important to persons now, (b) how they want to live their life, and (c) what supports will be necessary in the light of dementia diagnosis. The goal in these discussions would be to both explore and record, e.g., through a life story or a more formal statement specifics on the person's preferences.

Discussions of end-of-life issues in the view of the Summit are further conceptualised as a process of ongoing conversations over time, rather than one single formal event [32]. For so many people with an intellectual disability some of who have rarely been asked to make decisions about how to live their lives, receiving an invitation to attend an '*end of life meeting*' immediately following diagnosis of dementia would be confusing, possibly distressing and frightening, and for some meaningless. The Summit recommends that initiating end-of life discussions for people

with an intellectual disability and dementia be approached with sensitivity and within the context of incremental, supportive and educational long term planning.

Thinking Ahead

The Summit agreed that future care discussions should be guided by approaches that are intended to organize care in ways that help individuals lead full and fulfilling lives following diagnosis, yet respond to the progressive nature of dementia through additional discussions both at regular intervals and when transitions points and health care events occur, thus preventing and mitigating crisis situations [26, 33]. The Summit also emphasized that, like in the general population, there may be some individuals who do not wish to discuss any aspect of end-of-life care and that care teams, staff, and families are sensitive and respectful to their wishes [34].

The Summit also recognized that advanced and end-stage dementia and related end-of-life decision-making is an emotional and value laden time, complicated by close and often long standing relationships, yet is often marked by the limited ability to know and understand the wishes of the person or by the person's potentially not knowing what will transpire as they continue to decline. The Summit agreed that good practice is comprised of:

- Recognizing that end-of-life discussions are not a single event or a single discussion, or something that happens on the day of diagnosis.
- Discussing dementia care within the context of relationship-based approaches with those who know the person and understand his or her needs and preferences, habits and joys. In many cases this will be a family member and may include a partner or spouse with intellectual disability (or for some people with ID this may be paid carers).
- Supporting family and staff engagement in natural and meaningful conversations with persons with ID and advanced dementia regarding their previously expressed preferences gathering verbal and non-verbal clues on what continues to be important as they live their life with dementia.
- Utilizing life story strategies early in the dementia process (if not before) to develop a record of the person's preferences and wishes, likes and dislikes to inform later

discussions of care when the person is no longer able to participate in care decisions[35-36].

- Understanding the person's attempts at communication, responding appropriately when the person gives a clue, asking questions and listening to concerns or worries.
- Compiling a formal record of expressed wishes that is kept and ensuring that all person engaged in care know those wishes.

Examples of such frameworks and guidance for these discussions exist in many countries, such as a) "Zorgstandaard Dementie" in the Netherlands,[37] b) the UK's Gold Standard Framework,[38] c) New South Wales' 'State wide Framework for Palliative and EOL Care Service Provision',[39] d) the National Task Group' practice guidelines in the USA,[40] and e) the Irish Hospice Association's AFIRM,[41] and can be sourced for further reference. In the U.S., the Alzheimer's Association also has a useful end of life care decisions' booklet [42].

The Anticipatory Process

In accordance with the UK's NICE guidelines and quality standards on care of the dying adults in the last days of life [43] the Summit recommends that quality care for people with intellectual disability and advanced dementia include (a) monitoring of signs and symptoms that may signal the individual is transitioning into the terminal phase of dementia, including the dying phase; b) individualised care and genuine inclusion of those who matter to the person being present c) anticipatory prescribing and supportive care, and d) greater collaboration among intellectual disability services, hospitals, primary care, and specialist palliative care (also known as hospice care).

Monitoring signs and symptoms

Recognizing and assessing indications of when the person is at the terminal stage of dementia and in their last days of life is complex and is often confounded by other issues, such as acute episodes of delirium. Some people may present with ambiguous and conflicting signs and symptoms and will appear at times to be responding, showing signs of recovery which may

continue or be temporary. Signs and symptoms that may suggest that the person is in his or her last days of life include, increasing fatigue and diminished levels of consciousness, minimal oral intake, mottled skin and cool peripheries, altered respiratory rate, and finally, unconsciousness, noisy respiratory secretions, and Cheyne-Stokes breathing signalling the last hours of life. Assessment for change in signs and symptoms should occur frequently as these can often change quickly. It is critical that staff and family carers have access to professional medical and nursing support and that systems are in place to monitor symptoms and review changes in the person's condition to help determine if the person is nearing death, or stabilising. Working closely and in partnership with hospice and palliative care services will be important to help determine prognosis and measures now required to ensure a peaceful and dignified death, as well as offering reassurance and support to family and staff caregivers.

Individualised care

The Summit recognized the importance of individualised care and the genuine inclusion of those who matter to the person being present during this final and intimate phase of life. In particular, the person's family and staff carers should be given the opportunity to discuss, reflect, develop, and review the person's care plan considering the person's changing needs and condition. Quality reviews should also be undertaken to assure optimal end of life care. The Summit also proposed advanced dementia care be provided consistent with the person's needs and preferences, taking cognisance of their pre-existing level of ID and their engagement and involvement and control of in life decisions prior to dementia. This stage is but the conclusion of a meaningful life and families and paid staff should be supported in collecting stories, pictures, and valued possessions and in considering and expressing what mattered to the person. These activities help carers offer meaningful support for the person who is dying; and prepare for and offer essential support for later bereavement and the commemoration of a worthy life [44].

Anticipatory prescribing and symptom management

In the last few days of life, anticipating potential changes in health status, offering timely supportive care focused on symptom management, and alleviating psychosocial and spiritual distress, is essential in supporting a dignified death. It is important to understand that as the

person approaches his or her final days, there is likely to be changes in existing symptoms (such as seizures), as well as the emergence of new symptoms (such as agitation and breathing difficulties). In addition, difficulties in swallowing are likely to result in problems with taking essential medications orally. Prescribing medicines in anticipation, including the route of administration is important to avoid lapse in symptom control, avoiding unnecessary distress for the person (and at times for their family and staff carers). For example, alternative means of medication administration such as continuous subcutaneous infusions or rectal suppositories may be required to administer essential medication necessary to control seizures and other symptoms such as pain, breathlessness, respiratory secretions, or agitation.

The Summit endorsed the use of comfort measures of effective pain and symptom management, recommending the offering of a quiet, private environment that supports the intimate process of dying, providing for the person's spiritual preferences, and attending to oral and skin care [45].

Advanced dementia care recommendations

The Summit recommended that the choice of care strategies for persons with ID and advanced dementia be established by:

First in conjunction with **person-centred principles**, reviewing the values and preferences of the person with dementia,

Determining the **goals of care**, informed by facts and the person's life story; again with the focus upon the person

Determining what is in the '**best interest**' of the person considering the terminal nature of dementia.

Establishing the **intent** of treatment and the potential for beneficial outcomes versus burden.

Recognizing that **care decisions** arrived at by care teams must reflect the person's wishes, and be guided by family and friends when the person is no longer able to speak for him or herself.

Pursuing **care management** using a five-step process: 1) clarify the clinical situation, 2) establish primary goals of care, 3) present the treatment options and their risks and benefits, 4) weigh the options against values and preferences, and 5) provide additional and on-going support [46].

A case study illustrates the implementation of the Summit's recommended principles for quality care. Their translation for practice is summarized in Table 1 illustrating how each of five steps accommodates the presenting symptoms, the key factors that need to be considered, and the resulting care approaches.

Case History

Peter W. has Down syndrome and a moderate degree of intellectual disability. For most of his adult life he lived in a community home setting and was described as "a relatively independent man" who was competent in basic day-to-day activities of living. He attended a work placement five days a week, and went home to his family every weekend. He had limited verbal communication skills, using at best 1-2 word sentences; however, he made his needs known through facial expressions, body language, and gestures. He enjoyed relatively good health, had a history of hypothyroidism and visual impairment. At age 48 it was noted by staff and family that he was presenting with behaviour and personality changes, as well as memory problems and confusion. He was referred to a memory clinic specializing in dementia in persons with intellectual

disability and after a comprehensive diagnostic work-up he was diagnosed with Alzheimer's disease.

Peter's decline was progressive with marked changes in cognition and global day-to-day functioning and at the age of 53 years he had his first seizure. He was assessed on an annual basis using objective and informant based test instruments. At first assessment Peter scored 22 out of a maximum of 24 on the Test for Severe Impairment, 17 out of a maximum of 23 on the Daily Living Skills Questionnaire; and 11 out of a maximum of 22 on The Down Syndrome Mental Status Examination. Over a 7-year period Peter's cognitive and functional scores declined to zero.

As dementia entered the advanced stage, the community group home struggled despite their best efforts to give care with comfort or with safety and Peter was transferred to a setting with 24-hour staffing and nursing support. By this time, Peter had profound memory deficits, was completely dependent in all basic day to day activities of living, required the assistance of two staff members with bathing and toileting, and was presenting with increasingly complex medical needs (including recurrent chest infections and seizures). He had increasing difficulty with eating and drinking, and was put on a mechanically altered diet, finger foods, and nutritional supplements. Peter was reviewed on a regular basis by a multidisciplinary team that included the physician, nursing, social work, a speech and language therapist, dietician, and occupational therapist. His family was very involved in his care and his sister visited on a daily basis. The care team involved family and other carers, began planning with a focus on Peter's best interests, and, as may be seen in Table 1 considered the intent behind all treatments.

An end-of-life care plan reflecting Peter's past expressed wishes was developed in consultation with Peter's sister, his general practitioner, and members of the care team and was regularly reviewed. Specialist consultation and advice was sought from a palliative care consultant. Over next 14 months, Peter was increasingly holding food in his mouth, coughing, and spitting out the food, and there was a propensity to aspirate accompanied by frequent chest infections. Peter was subsequently admitted for 6 days to an acute hospital for the treatment of aspiration pneumonia (his third pneumonia in over a six-month period). Peter was treated with IV antibiotic, and IV fluids and oxygen therapy. The infection cleared, but Peter's overall condition

deteriorated and he was unable to eat or drink and swallow. An evaluation revealed advanced oral and pharyngeal dysphasia.

Peter was discharged on subcutaneous fluids which continued for a further 5 days. During this period comfort measures were continued, including use of a pressure mattress to promote comfort and prevent pressure areas. Peter appeared to enjoy iced flavoured mouth swabs and small tastes of ice cream given regularly on the tip of a special spoon, but overall there was minimal oral and caloric intake. A family meeting was held, the end-of-life plan was re-visited, palliative management was discussed, and it was agreed not to initiate tube feeding.

Five days post-hospital discharge, it was recommended to re-site Peter's subcutaneous line as he had become increasingly distressed, and had pulled at the line. It was also evident given facial expressions and audible whimpering that personal care activities were now experienced as burdensome. His sister and care team now concluded that Peter's prognosis was very poor and they did not wish to cause him any distress, and it was decided not to re-site the subcutaneous line. Comfort measures were continued, essential medication to control seizure activity and to relieve excessive secretions and agitation were administered via a continuous subcutaneous infusion. Peter died peacefully five days later surrounded by his family and carers and peers.

The case study illustrates that for people with Down syndrome, eating and swallowing difficulties and infections are a hallmark of advanced disease and impending death, supports findings noted in the general population and in other people with ID [26] (see Mitchell et al., 2009). In Peter's case, initial difficulties included holding food in his mouth, coughing, and spitting out food, and a propensity to aspirate. These difficulties were initially addressed using conservative measures. As dementia progressed these difficulties were confounded by the onset of acute infection and advanced dysphasia. A lack of swallowing and signs of distress caused the family and care team to consider a number of accepted practices, such as continuing to hydrate using subcutaneous fluids or inserting a long term feeding tube. In this case, the family decided against a feeding tube and then when evidence of distress and pulling at subcutaneous line was noted, it was decided that even this hydration intervention would not be re-sited. The route for the administration of essential medication to control seizures, manage secretion and terminal

agitation was done in a timely manner avoiding lapse in symptom control. Psychological, emotional and spiritual care supports were provided and the opportunity for Peter's family and friends to be with him during his final days was assured.

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The Summit noted that not every person with an intellectual disability lives within a facility and that dementia and advanced dementia may occur while living with family. Here the recommendation was that there be more investigation of such family caregiving situations, the uniqueness of care and of challenges experienced including challenges during transitions from home to facility which may be necessitated by levels of care needed in advanced dementia. Again early attention to understanding, documenting and implementing the individual's expressed preferences is critical. Achieving this will require earlier discussions with families about the risk for dementia, the value of documenting preferences and the creation of life stories as one potential tools. This is already recommended for the general population with dementia [47] and should be no less important for people with Down syndrome and other intellectual disabilities

Discussion

The Summit recognized that dying from a terminal condition, such as dementia resulting from Alzheimer's disease is rarely an acute event, but is one more likely to be of long duration, with multiple co-morbidities and events, with the potential to carry a high symptom burden. The seven-year findings presented here using a case history illustrate the clinical progression of Alzheimer's type dementia, and the transition to advanced dementia and death. As evidenced in Peter W.'s case, as dementia progresses and as life begins to draw to a close, difficult decisions will need to be made in relation to the continuance or discontinuance of various treatments and these are best managed by considering the *intent* of interventions, the *best interests* of the individual, whether interventions are *futile* and excessively burdensome, and by *involving the people who care* (family and staff) in all decisions. A key feature as illustrated in Peter W.'s care meetings, particularly early in the disease process, are educational opportunities with family and support staff to review the clinical course of dementia and what to expect in the later stages. This approach helps to provide the context for future discussions and decision-making as dementia progresses. Additionally, annual assessments using recommended test instruments,[39, 48-49]

help monitor progression and staging of the disease in the context of pre-existing intellectual impairment.

The Summit's consideration of this phase of dementia and life of persons with intellectual disability provided for the presentation of a series of recommendations that can further enhance quality care and promote quality of life. Included among the recommendations are (1) developing a practice guideline that can provide direction for staff activities in providing care at this latter stage of life and which incorporates the knowledge of body systems and the progression of losses in function, (2) ensuring that care workers who manage the immediate care of a person with advanced dementia are well trained in care practices and recognition of signs and symptoms of impending death, (3) incorporating a policy into services/ agency practices that recognizes the value of extended planning for end-of-life care preferences of the adult and his or her family and having the capacity within the care setting of palliative care to ease the distress prior to dying, and (4) developing partnerships with palliative and hospice care organizations, as well as with other groups that provide emotional supports to family members and other affected by the adults death.

The Summit participants noted that understanding the clinical course of advanced dementia, including prognosis and expected complications, combined with good multidisciplinary team and specialist palliative care guidance, will assist with complex decision-making for persons with an intellectual disability nearing end of life with advanced dementia, and help avoid potentially burdensome interventions where the evidence base for their use is unclear or not substantiated. A person-centred principle guided approach like the one illustrated in the case study can offer the most effective, systematic and caring strategy. End-of-life care is distressing for all persons involved, however, with supportive training and the institution of care practices based on knowledge of dementia and comorbidities, care practices can help raise the occurrence of a dignified death.

'In the past we did the best we could with what we knew, now that we know better, let's try and do better. (Adapted from Maya Angelou)[50].

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