

### Editor's key points

► Providing care for aging adults with intellectual and developmental disabilities is challenging, particularly with the onset of new or worsening chronic illnesses and the need for planning for the end of life. The family physician's role is multidimensional: providing medical care, advocating for additional services, and supporting and educating other professionals.

► This article provides guidance on understanding patients' cognitive ability; diagnosing and managing mental health issues; assessing patients' capacity to consent; discussing death and dying; creating advanced directives and living wills; and educating, advising, and coordinating with family, substitute decision makers, and other practitioners and caregivers.

### Points de repère du rédacteur

► La prestation des soins aux adultes vieillissants qui ont une déficience intellectuelle et développementale peut s'avérer compliquée, surtout lorsqu'une nouvelle maladie chronique apparaît ou qu'une maladie existante s'aggrave, et qu'il faut planifier en vue de la fin de vie. Le rôle du médecin de famille compte plusieurs facettes: dispenser des soins médicaux, plaider en faveur de services supplémentaires, et appuyer et former les autres professionnels.

► Le présent article conseille les médecins sur la façon de comprendre les capacités cognitives des patients; de poser un diagnostic de trouble de santé mentale et de le prendre en charge; d'évaluer la capacité du patient à donner son consentement; de discuter de la mort et du processus de la mort; de produire des directives de planification des soins et un testament de vie; et d'éduquer et de conseiller la famille, les mandataires et les autres professionnels et soignants, et d'assurer la coordination.

# Approach to providing care for aging adults with intellectual and developmental disabilities

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## Abstract

**Objective** To provide an approach to caring for aging adults with intellectual and developmental disabilities (IDD) in the context of the onset of new or worsening chronic illnesses and the need for planning for the end of life.

**Sources of information** A MEDLINE search identified few review articles in the past 10 years. This review builds on relevant articles and the experiences of the author and colleagues working with aging adults with IDD and their families, physicians, and other caregivers.

**Main message** To provide care to this patient group, physicians must understand the diverse cognitive abilities of adults with IDD; the risk factors for physical and mental illnesses; concerns related to diagnostic overshadowing; and the need for coordinating individual care plans for those with serious and terminal illnesses.

**Conclusion** Primary care physicians can provide and coordinate appropriate care for patients with IDD as they face the health challenges associated with aging and dying. Being aware of patients' baseline cognitive abilities and decision-making skills, as well as changes in cognitive abilities associated with aging and complexity of illness, will help determine patients' capacity to consent, identify appropriate treatment choices, and guide coordination of care. Further research and consensus statements are needed to guide best practices based on the Canadian experience and to allow continuing development of caring, professional, and competent providers to support aging adults with all levels of IDD.

## Approche des soins aux adultes vieillissants qui ont une déficience intellectuelle et développementale

### Résumé

**Objectif** Fournir une approche pour les soins aux adultes vieillissants qui ont une déficience intellectuelle et développementale (DID) dans le contexte de l'apparition d'une nouvelle maladie chronique ou de l'aggravation d'une maladie existante, et du besoin de planifier en vue de la fin de vie.

**Sources de l'information** Une recherche sur MEDLINE a relevé peu de revues de synthèse ayant été publiées dans les 10 dernières années. Cette revue s'appuie sur des articles pertinents ainsi que sur l'expérience de l'auteur et de ses collègues dans le cadre de leur travail auprès d'adultes vieillissants qui ont une DID et de leur famille, de leurs médecins et d'autres soignants.

**Message principal** Pour dispenser des soins à ce groupe de patients, les médecins doivent comprendre les différentes capacités cognitives des adultes ayant une DID; les facteurs de risque de maladie mentale et physique; les préoccupations en matière de dissimulation diagnostique; et le besoin de coordonner les plans individuels de soins chez les patients atteints d'une maladie grave ou terminale.

**Conclusion** Les médecins de première ligne peuvent dispenser et coordonner les soins aux patients ayant une DID lorsqu'ils font face aux problèmes de santé liés au vieillissement et à la mort. Le fait de connaître les capacités cognitives initiales du patient et ses capacités de prendre des décisions, de même que les variations des capacités cognitives associées au vieillissement et à la complexité de la maladie aidera le médecin à déterminer si le patient

est capable de donner son consentement, d'identifier les choix thérapeutiques appropriés et de guider la coordination des soins. Plus de recherche et d'énoncés de consensus sont nécessaires pour orienter les pratiques exemplaires fondées sur l'expérience canadienne et pour former des fournisseurs de soins attentifs, professionnels et compétents qui soutiennent les adultes vieillissants ayant une DID de tout niveau.

## Case description

Philip, a 67-year-old man with moderate intellectual and developmental disability (IDD), presents with fatigue, cough, and weight loss. He has a 50-pack-year history of smoking. He is accompanied by a developmental services worker who reports that Philip lives alone and makes his own medical decisions. He lived in an institution until 5 years ago. A staff member from the agency assisting with his care comes to his apartment once a day. Philip does not want to go to the hospital "because people die there."

Providing care for aging adults with IDD is challenging, particularly with the onset of new or worsening chronic illnesses and the need for planning for the end of life. The family physician's role is multidimensional: providing medical care, advocating for additional services, and supporting and educating other professionals.

## Sources of information

Canadian best practices guidelines have been developed for the care of this vulnerable population<sup>1,2</sup> but they do not specifically address the challenges of an aging population. A MEDLINE search for articles in English published in the past 10 years using key words including *aging*, *intellectual disability*, *patient care*, *nursing care*, *delivery of health care*, *end-of-life*, and *capacity to consent*, identified no specific guidelines for the care of aging adults with IDD. Articles from Great Britain, the Netherlands, the United States, and other countries reported on case studies, children's issues, syndrome-specific issues, and services available to this group that are different from those available in Canada. Publications did not routinely specify the cognitive abilities of patients, making it challenging to generalize recommendations to this diverse population. Lunsky and colleagues emphasize the need for more Canadian data based on the health care needs of aging IDD populations and the Canadian health care system.<sup>3</sup>

This clinical review is based on the experiences of the author and colleagues working with aging adults with IDD and their families, physicians, and caregivers, as well as relevant material from the literature search.

## Main message

To provide care to this cohort, doctors must understand the following:

- the implications of the diverse cognitive abilities of adults with IDD;

- risk factors for physical illnesses, including advancing age, smoking history, nulliparous status;
- risk factors for mental illnesses, including losses and family history;
- concerns related to diagnostic overshadowing and multidisciplinary care for those with serious and terminal illnesses;
- changing capacity to consent to treatment owing to age-related cognitive deterioration or increasing complexity of treatment choices; and
- use of advanced directives to guide treatment and terminal care options.

**Understanding cognitive ability.** Cognitive ability among people identified as having IDD can vary considerably, so it does patients a disservice to make broad recommendations for all. Adults with mild disability live independently and might not have had their IDD formally identified. Patients with mild or moderate intellectual delay can understand new information and make decisions at a more complex level than those with severe or profound IDD. **Table 1** compares mental age, IQ, and functional descriptions of cognitive disabilities.<sup>4</sup>

A person with mild IDD and good verbal abilities who lives alone might present the greatest management challenge. He or she might have refused support owing to stigma. However, increasing complexity in illnesses might result in an inability to understand risks and benefits or to follow a treatment plan. The physician might suspect mild IDD only after challenges in health provision are noted over time.

Identify a person trusted by the patient, such as a spouse, friend, or adult child, to accompany the patient with mild IDD to appointments and procedures to help with plans and report challenges. Have the patient sign consent forms for communication with this individual.

**Diagnosing physical illnesses.** Read and Thompson-Hill report that individuals with IDD receive fewer screening tests and investigations and are 4 times more likely to die of treatable illness than those in the general population.<sup>5</sup> The Canadian Health Watch Tables for adults with intellectual delay<sup>2</sup> are useful resources when choosing screening tests in patients with IDD; however, there is evidence that primary care guidelines developed for specific syndromes associated with IDD are not applied consistently.<sup>6</sup>

A developmental approach to understanding symptom presentation and diagnostic issues helps practitioners remember that, although age is a risk factor for many illnesses, adults with IDD might present with atypical symptoms. Chronic or acute pain or a new-onset medical illness might present with developmental-age-appropriate signs of distress. Attribution of symptoms of distress to the IDD, known as *diagnostic overshadowing*, can result in delayed recognition of new or worsening physical illness.

Clinical suspicion of new illness should be based on the patient's chronologic age and other risk factors; cognitive level will contextualize symptom presentation.

**Table 1. Comparison of cognitive level, IQ, mental age, Piagetian stage, and communication skills in patients with cognitive disabilities**

COGNITIVE LEVEL	IQ	MENTAL AGE, Y	PIAGETIAN STAGE	COMMUNICATION SKILLS
Borderline	70 to 84	12 to 16	Formal operations	Read and write at a grade 2 to 8 level
Mild	55 to 69	8 to 12	Concrete operations to formal operations	Read and write simple phrases, ability to follow simple instructions using prepositions
Moderate	43 to 54	5 to 8	Preoperational to concrete operations	Minimal sight vocabulary, understands 2-step commands
Severe	Hard to test	2 to 5	Preoperational	Telegraphic speech, understands 1-step commands
Profound	Hard to test	Infancy to 2	Sensorimotor	Orients to speech, small vocabulary

Adapted from Lougheed.<sup>4</sup>

Encourage all patients with IDD to identify and describe their own concerns. The person accompanying the patient can help describe and clarify symptoms but might bring his or her own biases into the description. This can be overcome with explanation and gentle rephrasing of questions without criticism.

Caregivers of those with severe and profound IDD should be questioned using a structured medical interview.

**Diagnosing mental health issues.** Identifying new or recurrent psychiatric illness in aging adults with intellectual delay is difficult. Anxiety and depression are common in adults with IDD<sup>7</sup> and can present with episodes of aggression and avoidance. Making an accurate diagnosis is confounded in patients with moderate or severe IDD who have difficulty recognizing or reporting symptoms or understanding questions; they might attempt to hide symptoms to appear “normal,” or there might be unusual presentations of symptoms.<sup>8</sup> Staff caregivers might identify new-onset symptoms as “behaviours,” attributing challenging episodes that are reactions to pain or fatigue to willfulness or lack of cooperation.

Physical illnesses, such as hypothyroidism, pain from occult fractures, urinary tract infections, or constipation should be ruled out while mental health issues are clarified.

Standard protocols for investigating and treating diagnosed psychiatric illness should be used as per geriatric population guidelines (eg, that by Mulsant et al).<sup>9</sup>

Distinguishing between depression, delirium, and new-onset dementia is compounded in adults with IDD by diagnostic overshadowing. This is especially challenging in aging patients with severe or profound IDD who might be at greater risk of polypharmacy and its related side effects.<sup>7</sup>

Review and reduce all medications that might contribute to delirium.

Treat for depression, owing to its better prognosis relative to dementia. This is particularly important in adults with trisomy 21, for whom depression is common.<sup>2</sup>

Depression and anxiety might present with symptoms of irritability, hitting, or slowness in daily activities. Dementia should be considered only when other treatable causes of confusion, personality change, or memory problems are ruled out.

Despite limited research looking systematically at precipitants of mood or anxiety disorders in adults with IDD, clinical experience can guide family physicians:

- Recent loss of family, caregivers, or friends; poor physical health; reduced ability to socialize; family history; or a recent move can be triggers or factors.<sup>7</sup>
- Caregiver neglect or abuse, including prescription or over-the-counter medication misuse, and caregiver burnout or illness need to be considered.
- Alcohol or substance abuse by patients with mild IDD needs to be considered, and collateral information should be obtained from a spouse, friend, or other family member.
- Individuals from nondominant cultural backgrounds (eg, refugees or recent immigrants) might be at more risk of mental health issues and might have difficulty finding resources.

Family meetings coordinated by the family physician can help identify resources and educate caregivers and family members about planning and support needs.<sup>10</sup>

Biopsychosocial treatment planning models should guide treatment.

**Assessing capacity to consent.** Capacity to consent varies both with older adults’ baseline cognitive ability and with the increasing complexity of necessary treatment options (Table 1).<sup>4</sup>

Discuss choice of a substitute decision maker (SDM) with adults with mild or moderate IDD. Know about relatives and friends who are possible options and identify a way to contact them. Include all relevant documents including consents in the patient’s chart.

According to *Tools for the Primary Care of People with Developmental Disabilities*,<sup>2</sup> capacity is the mental ability to make a particular decision at a particular time. Physicians must follow appropriate legal and ethical guidelines to determine consent.

To assess capacity to consent to a specific treatment, ask questions related to the risks and benefits of the treatment and of no treatment. For instance, “Do you know why you are in the hospital now? Do you know what might happen if you have or don’t have this treatment?”<sup>2</sup> Date and record the findings in the chart.

If the patient is found to be incapable by the treating physician, the physician must identify the legal SDM and obtain

consent from that person. If consent givers who are equally ranked disagree (for instance, 2 siblings), mediation by the physician can be helpful. Otherwise, the physician should contact the hospital lawyer or the provincial Office of the Public Guardian and Trustee for advice.

Older adults with mild IDD might be able to continue to make treatment decisions even as the complexity of their illness and treatment increase. However, they benefit from having a family member or friend present to help them cope with challenging news or to discuss treatment options. Adults with moderate intellectual delay usually are accompanied by family members or staff from a developmental support agency. According to provincial consent and capacity law, paid staff caregivers cannot give consent on behalf of their charges, even if there are written directives from the SDM. This point is poorly understood by developmental agency staff.

After interviewing the patient, decide who will be providing consent for treatment—the patient or an SDM. Always contact the legal SDM for review of diagnosis and treatment recommendations and for planning. This can be done by telephone and documented in the chart. Note details about proposed medications and dosage ranges, other treatments, and possible side effects.

As details vary among the provinces and territories, family physicians with limited experience applying legislation around capacity to consent and identification of SDMs should seek advice from doctors working with the elderly, where changing capacity issues are common, or from hospital-based lawyers.

**Discussing death and dying.** Wagemans and colleagues reviewed medical end-of-life decisions in adults with a range of IDD in a residential care setting in the Netherlands. Families were involved in decision making in half of deaths, but patients were never asked for their opinions.<sup>11</sup> McEvoy et al reported that a sample of 30 adults had a limited understanding of the basic biological components of death, and that the level of understanding was associated with the level of IDD.<sup>12</sup> Tuffrey-Wijne and Rose reported that staff caregivers found it difficult to support individuals with IDD around death and dying owing to their own concerns as well as organizational culture issues.<sup>13</sup>

Reviewing levels of cognitive ability and mental age, as presented previously,<sup>4</sup> might help physicians develop their skills and increase their comfort in talking about illness and death with adults with IDD and their families and caregivers.

**Creating advanced directives and living wills.** Adults with IDD should not be excluded from the important end-of-life step of developing advanced directives and living wills. Adults with mild disability should be supported in planning care choices for serious or terminal illnesses. For patients who are not capable of independent decisions, SDMs should be consulted when possible.

Advanced directives can include identifying desired levels of invasive investigation or resuscitation orders; guidance of treatment in palliative situations; care for

dementia; and when or if to transfer a patient to a long-term care facility. Living wills often include identification of favourite activities and music, sleep habits, favourite clothes, and communication needs.

Wagemans and colleagues write that, for physicians caring for patients with IDD, the planning of do-not-resuscitate orders is complex, with quality-of-life issues causing conflict between physicians and relatives.<sup>14</sup> They encourage development of advanced care planning documents. Stein and Kerwin recommend more dialogue, research to inform palliative care resources, and new models for providing care for adults with IDD.<sup>15</sup> Based on a literature review, Clarke and colleagues reported that treatment decisions involving individuals who lack capacity to make their own decisions tend to be based on factors such as improving quality of life, prolonging life, and patient wishes.<sup>16</sup> In a consensus statement, McCallion and an international multidisciplinary group define the state of advanced dementia in adults with IDD, and recommend involving patients early in their advanced care planning.<sup>10</sup> Vrijmoeth et al report that physician referral of patients to palliative care is influenced by diagnostic uncertainties in people with IDD, patients' communication skills, and the need for reliance on caregivers.<sup>17</sup>

Interview patients to assess their understanding of end-of-life issues and their capacity to make difficult decisions, and clarify wishes, even if patients are found not to be competent. Time-consuming discussions need to be booked appropriately. Use plain language appropriate for the patient's comprehension and cognitive skills, and repeat information.

Direct the staff members of the group home to explore what their agency can provide for palliative care or support in hospital.

Encourage older SDMs, such as parents, to identify a successor, as the provincial Public Guardian and Trustee becomes the default decision maker if no one else is identified. Encourage SDMs to discuss, identify, and record their wishes for the care of their ward and ask them what the adult with IDD would want for him or herself.<sup>10</sup>

As of 2017, Canadians have access to medical assistance in dying if they have an illness likely to result in death in the near future. Adults with IDD are eligible to apply if they meet the criteria for capability of consenting to or refusing personal care and making medical decisions. An SDM cannot request medical assistance in dying for an incapable person but can provide end-of-life care decisions.

Advanced directives are not legally binding, but they provide guidance for the physician, the care team, and family members. Keep copies at the office and the patient's residence, and review annually or when the patient's situation changes.

**Educating, advising, and coordinating.** Besides providing medical care, the family physician is an advocate, advisor, and health care system interpreter for the patient, family, SDM, and developmental sector agencies that provide support. This role includes the following:

- helping the patient (and SDM) understand the diagnosis and the pros and cons of treatment, watchful waiting, or no treatment;

- supporting and encouraging caregiving staff and identifying and addressing lack of knowledge, skills, and resources for staff and families<sup>13,18</sup>;
- coordinating services across agencies covering housing, financial support, home care, and medical requirements; and
- advocating for in-hospital agency support for a patient who might be disruptive if hospitalized, for ancillary resources in serious illness, and for palliative care and dying at home, if appropriate.

Each adult with IDD requires individualized planning with a team approach. Written care plans should be reviewed as illness progresses. Physicians need to advocate within the system for increased support in the community and in the hospital for patients living alone, with family, or in a developmental sector-supported setting.

### Case resolution

Philip's physician assessed him in the office for capacity to consent and identified him as not being capable of consenting or refusing treatment for this illness. Philip was able to identify a cousin he liked, and the agency responsible for his care helped contact this nearest next of kin in another province. She agreed, with support and education, to provide consent for treatment for emphysema and later for lung cancer.

The agency agreed to provide Philip with support to attend consultation visits, increased its support in the home after hospitalization, and, 2 years later, arranged for placement in a group home where smoking was not permitted. Meetings coordinated by the physician with Philip, his SDM (by telephone), and the agency identified advance directives and a living will. Philip died peacefully in his new home with caring support.

### Conclusion

Primary care physicians can provide and coordinate appropriate care for patients with IDD facing the health challenges associated with aging and dying. Being aware of baseline cognitive abilities and decision-making skills, as well as changes in cognitive abilities associated with aging and complexity of illness, will help outline appropriate treatment and coordinate care plans with patients, family members, appropriate agency services, and other health care professionals. Interventions will reflect the wishes of the patient or the person providing substitute consent. In addition, use of available community support will ensure the best possible care for these complex patients at the end of life.

Further research or consensus statements are needed to guide best practices based on the Canadian experience and to allow continuing development of caring, professional, and competent providers to support aging adults with all levels of IDD.



Dr Lougheed is a practising psychiatrist and Assistant Professor at the University of Ottawa in Ontario.

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#### Competing interests

None declared

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#### References

1. Sullivan WF, Diepstra H, Heng J, Ally S, Bradley E, Casson I, et al. Primary care of adults with intellectual and developmental disabilities. 2018 Canadian consensus guidelines. *Can Fam Physician* 2018;64:254-79 (Eng), e137-66 (Fr).
2. Sullivan WF, Developmental Disabilities Primary Care Initiative Scientific and Editorial Staff, editors. *Tools for the primary care of people with developmental disabilities*. Toronto, ON: Surrey Place, MUMS Guidelines Clearinghouse; 2011.
3. Lunsy Y, Balogh RS, Cobigo V, Isaacs B, Lin E, Ouellette-Kuntz HM. Primary care of adults with developmental disabilities in Ontario. *Healthc Q* 2014;17(3):11-3.
4. Lougheed DC. Considerations in working with individuals who are homeless, have intellectual delay and mental illness. In: Clark C, editor. *Homelessness: prevalence, impact of social factors and mental health challenges*. Hauppauge, NY: Nova Science Publishers Inc; 2014. p. 191-201.
5. Read S, Thompson-Hill J. Palliative care nursing in relation to people with intellectual disabilities. *Br J Nursing* 2008;17(8):506-10.
6. Jensen KM, Taylor LC, Davis MM. Primary care for adults with Down syndrome: adherence to preventive healthcare recommendations. *J Intellect Disabil Res* 2013;57(5):409-21.
7. Tasse MJ, Macho PN, Havercamp SM, Benson BA, Allain DC, Manickam K, et al. Psychiatric conditions prevalent among adults with down syndrome. *J Policy Pract Intellect Disabil* 2016;13(2):173-80. Epub 2016 May 11.
8. Fletcher RJ, Barnhill J, Cooper SA, editors. *DM-ID 2. Diagnostic manual—intellectual disability: a textbook of diagnosis of mental disorders in persons with intellectual disability*. Kingston, NY: NADD; 2016.
9. Mulsant BH, Blumberger DM, Ismail Z, Rabheru K, Rapoport MJ. A systematic approach to the pharmacotherapy of geriatric major depression. *Clin Geriatr Med* 2014;30(3):517-34. Epub 2014 Jun 14.
10. McCallion P, Hogan M, Santos F, McCarron M, Service K, Stemp S, et al. Consensus statement of the International Summit on Intellectual Disability and Dementia related to end-of-life care in advanced dementia. *J Appl Res Intellect Disabil* 2017;30(6):1160-4. Epub 2017 May 9.
11. Wagemans A, van Schroyen Lantman-de-Valk H, Tuffrey-Wijne I, Widdershoven G, Curfs L. End-of-life decisions: an important theme in the care for people with intellectual disabilities. *J Intellect Disabil Res* 2010;54(6):516-24. Epub 2010 Mar 26.
12. McEvoy J, Treacy B, Quigley J. A matter of life and death: knowledge about the body and concept of death in adults with intellectual disabilities. *J Intellect Disabil Res* 2017;61(1):89-98. Epub 2016 Nov 11.
13. Tuffrey-Wijne I, Rose T. Investigating the factors that affect the communication of death-related bad news to people with intellectual disabilities by staff in residential and supported living services: an interview study. *J Intellect Disabil Res* 2017;6(8):727-36. Epub 2017 Apr 7.
14. Wagemans AM, van Schojenstein Lantman-de Valk HM, Proot IM, Bressers AM, Metsemakers J, Tuffrey-Wijne I, et al. Do-not-attempt-resuscitation orders for people with intellectual disabilities: dilemmas and uncertainties for ID physicians and trainees. The importance of the deliberation process. *J Intellect Disabil Res* 2017;61(3):245-54. Epub 2016 Aug 26.
15. Stein GL, Kerwin J. Disability perspectives on health care planning and decision making. *J Palliative Med* 2010;13(9):1059-64.
16. Clarke G, Harrison K, Holland A, Kuhn I, Barklay S. How are treatment decisions made about artificial nutrition for individuals at risk of lacking capacity? A systematic literature review. *PLoS One* 2013;8(4):e61475.
17. Vrijmoeth C, Barten P, Assendelft W, Christians M, Festen D, Tonino M, et al. Physicians' identification of the need for palliative care in people with intellectual disabilities. *Res Devel Disabil* 2016;59:55-64.
18. Tuffrey-Wijne I, Rose T, Grant R, Wijne A. Communicating about death and dying: developing training for staff working in services for people with intellectual disabilities. *J Appl Res Intellect Disabil* 2017;30(6):1099-110. Epub 2017 Jul 5.

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