

Family Medicine Curriculum Resource: Adults with Developmental Disabilities



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Family Medicine Curriculum Resource: Adults with Developmental Disabilities

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Developmental Disabilities in Adults A Curriculum Resource for Family Medicine Residency Programs*

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Family Medicine Curriculum Resource: Adults with Developmental Disabilities

Part 1: Competencies



Family Medicine Curriculum Resource: Adults with Developmental Disabilities

Part 1 – Competencies*

A resident competent to enter independent family practice...

1. Ethical Considerations

- a) Demonstrates respect and empathy for patients with developmental disabilities as individuals (e.g., in the way the resident includes the person with DD in the clinical encounter rather than speaking about him/her to the caregiver).
- b) Facilitates informed consent in persons with partial decision-making capacity (e.g., by asking if they would normally ask for help in making similar decisions).
- c) Identifies and supports substitute decision-makers (e.g., in using best practices as a guide to decision-making).

2. Communication and Access to Care

- a) Communicates effectively, applying communication methods in accord with the patient's individual expressive and receptive communication capabilities (e.g., simple language, writing and pictures, taking extra time).
- b) Adjusts the clinical environment (e.g., removes barriers to physical exams) for patient comfort, taking into account sensory integration issues.
- c) Facilitates access to the family practice team, consultants, hospital services (e.g., planning visits to imaging departments, consulting mental health services for a person with the dual diagnosis of mental health or behavioural issues plus an intellectual disability) and community resources (e.g., completing disability applications, directing to developmental services in the social and educational sectors).

3. Etiology of DD and Level of Function – Required Baseline Information

- a) Uses etiologic information (e.g., a diagnosed genetic syndrome like Down syndrome) for anticipatory and preventive care and for acute presentations. When etiologic information is not available, assesses the need for further investigation (e.g., referral for genetics assessment or neuroimaging).
- b) Uses (or obtains, if necessary) up-to-date assessments of intellectual and functional abilities (e.g., psychological, educational or vocational assessments) to determine appropriate expectations in clinical encounters and the adequacy of supports.

4. Conditions Commonly Associated with DD and Atypical Presentations of Serious Illnesses

- a) In generating differential diagnoses and management plans, demonstrates knowledge of the frequency, preventability, and treatability of various health issues specific to adults with DD (e.g., the most common cause of death is respiratory disease; common conditions associated with DD are autism, epilepsy, cerebral palsy, mental health problems; the importance to health and behaviour of transitions between life stages, such as adolescence to adulthood, or social situations, such as family home to supported living in the community).
- b) Recognizes atypical presentations of serious illness (e.g., behaviour change as a presentation of reflux esophagitis) and manages appropriately.

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Family Medicine Curriculum Resource: Adults with Developmental Disabilities

Part 2: Primer for Family Physicians

Based on
Primary care of adults with developmental disabilities:
Canadian consensus guidelines



Family Medicine Curriculum Resource: Adults with Developmental Disabilities

Part 2 – Primer for Family Physicians*

Based on Primary care of adults with developmental disabilities: Canadian consensus guidelines by Sullivan et al, Can Fam Physician 2011 [1,2] and the Developmental Disabilities Primary Care Initiative, Tools for the Primary Care of People with Developmental Disabilities

A. Introduction

Definition of DD

The terms developmental disabilities (DD) or intellectual disabilities are used to refer to a range of conditions in which lifelong limitations in intellectual functioning and conceptual, social and practical skills are noticeable before age 18 years.

Etiologies or conditions associated with DD

- Some persons with DD have diagnosed syndromes such as Down syndrome or fragile X; some fall under the umbrella term fetal alcohol spectrum disorder; some have associated conditions such as autism spectrum disorder or cerebral palsy.
- Many persons with DD do not have a diagnosed etiology or specific associated disorder. Where the etiology is known or suspected, it can be considered to be genetic and/or environmental in origin. The latter could have occurred in prenatal, perinatal or postnatal periods; for instance, prenatal infections, brain injury at birth and childhood traumatic brain injuries or CNS infections could be implicated.

How prevalent is DD?

As a demographic category, people with DD make up 1 to 3% of the population, so a general practice of 1,500 patients could be expected to include 15 to 45 such patients, who likely have a variety of vulnerabilities including poorer health status, barriers to access to health care and lower socio-economic status.

Why is DD important to FPs?

Canadian family physicians have an increasing and valuable role to play in serving this population, in particular in the care of adults with developmental disabilities, for whom, unlike children, there is no other generalist medical care provider group to meet patient and family expectations for care in the community and close to home.

B. Guidelines and Tools

Two documents provide Guidelines ([English](#) and [French](#)) [1,2] and Tools ([English](#) and [French](#)) for Canadian family physicians. Using them provides a basis for a Family Medicine curriculum in Developmental Disabilities.

The **Guidelines** are in three areas:

- i. Important general issues, e.g., establishing etiology and level of adaptive functioning, atypical presentations of pain and distress, risks of polypharmacy, abuse and neglect, appropriate communication techniques, advance care planning at times of life transitions and interdisciplinary care.
- ii. Physical health guidelines, highlighting preventive care and common morbidities.
- iii. Behavioural and mental health guidelines, emphasizing that, while psychiatric conditions are more common in this population, behavioural “problems” are more often a method of communication about social and environmental issues.

The **Tools**, currently about 25 in number, were designed with user input to help family physicians implement the Guidelines in practice. Most of the Tools are explained and linked in the next section of this Primer (“Pearls and Resources”), but two are worth noting here:

- [Cumulative Patient Profile](#)
- Preventive Care Checklist (forms for [males](#) and [females](#))

Because there is RCT evidence [3,4,5] for the benefit of periodic (probably annual [6]) comprehensive health exams for persons with DD, medical record forms/templates for a [Cumulative Patient Profile](#) and Preventive Care Checklist [[male](#) and [female](#) forms] have been developed, based on forms familiar to Canadian family physicians but adapted for DD. Whether used “as is” in a paper patient chart or integrated with existing electronic medical records, these forms can provide a structure for a patient encounter for comprehensive health review that follows the Guidelines (see Guideline 1, Appendix 1).

C. “Pearls” and Curriculum Resources for Key Features/Competencies

The Guidelines and Tools are the basis for the following “pearls” and resources, which have been selected to support the **four clinical competencies** for residents (see Part 1 of this Curriculum Resource for a summary of these competencies).

1. Ethical Behaviour

Respect and empathy

- Respect and empathy are demonstrated in the way the resident speaks and interacts with the patient and caregiver. The extent and skill with which the resident assesses and adapts to the communication level of the patient is a specific competence with respect to DD.

Consent and Substitute Decision-Makers (see Guidelines 7, 8 in Appendix 1)

- Residents should initiate the consent process for a person with DD when a new treatment or a change in treatment is proposed (unless it had been accepted through a previously agreed-to plan of care) or an assessment/investigation is proposed (especially if invasive).
- Half of persons with mild intellectual disability have capacity for informed consent (or withholding consent) for most medical procedures; 18% with moderate intellectual disability and none of those with severe or profound intellectual disability would have such capacity.
- A [tool](#) is available to assess capacity in persons with DD.
- Assessing capacity to consent and seeking consent requires supporting the patient by adapting the level and means of talking about an investigation or treatment to the patient’s level of intellectual and adaptive functioning. An adult with DD assessed as incapable of some aspects of decision-making (e.g., understanding or judging consequences) might still be able to convey, through verbal or other means, perspectives that can inform the judgment of a substitute decision-maker.
- The hierarchy of substitute decision-makers is identified by provincial legislation. In Ontario, and possibly in other provinces, paid caregivers, such as group home staff, cannot legally consent to or refuse treatment on behalf of a patient, but may be very helpful in providing guidance about communicating with patients and advice about obtaining assent or cooperation. In addition to the patient’s perspective in pursuing or forgoing any health care intervention, “best practices” may be the appropriate standard for decision-making, because, unlike people who lose the capacity for consent later in life, persons with DD

may not have reached the stage of development where their advance directives could be made known to the substitute decision-maker (Guideline 8, Appendix 1).

2. Communication and Access to Care

Communication (see Guideline 7, Appendix 1)

- Techniques to facilitate communication are identified in a [list of practical tips](#).
- It will generally take more time to communicate and establish rapport with persons with DD.

An assessment of language skills helps to choose the level of language to use. An awareness of communication strategies useful for persons with various levels of intellectual disability (mild, moderate, severe or profound) can be helpful. The [Adaptive Functioning and Communication Associated with Different Levels of Developmental Disabilities](#) is a quick guide to help practitioners assess an individual.

- Many people with DD have stronger receptive (understanding) communication skills than expressive skills. Conversely, the person's expressive speech may sometimes give an impression of better comprehension than is actually the case. It is important to check the person's understanding. Involving caregivers who know the person well may help better understand his/her subjective experiences. However, continue to focus communication efforts on the person rather than his/her caregiver.
- Behaviour is a form of communication; distress signs and behaviours are unique to the individual and may not be specific to a particular cause.
- Vision and hearing problems are under-diagnosed.

Other Aspects of Access to Care (see Guideline 9, Appendix 1)

- Thinking ahead about [office and waiting room environments and routines](#) in relation to the needs of the person with DD is important. Consider a home visit; mechanical lifts or assistants may be needed to facilitate positioning of the patient for a proper physical exam. Desensitization through story/picture books or practice simulations for venipuncture, imaging or other procedures such as Pap tests may be necessary (see the Notes to the Guidelines in the Appendix to this Primer for specific resources).
- The whole health team may be usefully involved in a planned way. A Caregiver Health Assessment form ([English](#) and [French](#)), mailed to the patient/caregiver in advance of a periodic comprehensive health review, can provide background

information and save time for the patient in the office. In such a review, individual tasks can be assigned to different staff and spread over time and follow-up visits to minimize the load on individuals. A form suitable to help patients and caregivers collect background information in advance of more incidental visits is also available: the [Today's Visit](#) form. Forms for recording [seizures](#), [sleep](#), [bowel movements](#), [weight](#) and [menses](#) are available.

- Asking about the educational, financial and social supports being used by patients with DD is important background information in assessing determinants of health (see [Community Resources in Ontario](#)).
- A checklist is available to help family physicians identify areas for advocacy on behalf of patients ([Advocacy Role of Family Physician/Advanced Practice Nurse](#)). These resources may be especially important in anticipation of health status changes caused by stressful transitions from school services to community services, from parental home to other supported living situations, and with aging.

3. Etiology and Level of Function – Baseline Information for Clinical Encounters

Etiology (see Guideline 2, Appendix 1)

- Etiology or associated conditions are useful to establish, whenever possible, to help inform anticipatory, preventive and acute care specific to those conditions or syndromes. Brief guidelines, designed for family practice and listing such specific issues by body system, are available for several of the most common conditions ([Down syndrome](#), [fragile X](#), [Prader-Willi](#), [Smith-Magenis](#), [22q11.2del fetal alcohol spectrum disorder](#) and [Williams syndrome](#); cerebral palsy and autism spectrum disorder are soon to be published). Peer-reviewed disease descriptions with management summaries – [Gene Reviews](#) – are available.
- Advances in genetic knowledge continue to enhance the detection of etiology, so periodic reassessment is appropriate for persons whose previous testing was inconclusive. Reasons to seek a genetic assessment include identifying an etiology that might have health management consequences for other family members (e.g., fragile X) to identify risk of recurrence within a family and to provide reassurance to patients and their families by identifying a cause. The [Genetic Assessment FAQs](#) tool provides answers to questions relating to making referrals to a genetics centre. Contact information for [Genetic Centres in Canada](#) is available.

Adaptive Functioning (see Guideline 3, Appendix 1)

- Adaptive functioning or adaptive behaviour refers to the skills (conceptual, social and practical) that a person with DD has to handle the common demands

of everyday life. Understanding the intellectual abilities and adaptive functioning of persons with DD sets the stage for productive clinical encounters and guides assessment, treatment and support plans. The tool, [Adaptive Functioning and Communication Associated with Different Levels of Developmental Disabilities](#), is a quick guide to help practitioners assess an individual.

- Although intellectual abilities described in terms of IQ or age equivalence inform the assessment of adaptive functioning, there is not necessarily a direct correlation between the two, i.e., an individual with high functioning autism may have a low level of adaptive functioning due to socio-cognitive difficulties without having an intellectual disability.
- Adaptive functioning can decline or improve in some adults with DD. A current assessment of intellectual and adaptive functioning helps to determine necessary care (e.g., ability to manage medications) and supports (e.g., need for support in attending appointments). It establishes a baseline for future assessment, which can be important in assessing behavioral problems or dementia.
 - Records of previous assessments may be available from school records.
 - Current assessments can be sought, e.g., from psychologists or occupational therapists and, in terms of communication, from speech language pathologists. DD service agencies, (in Ontario, the [Community Networks of Specialized Care](#) or [Developmental Services Ontario](#) or rehabilitation, vocational or mental health programs with such staff resources, may be able to provide such an assessment without cost to the person. The [Psychological Assessment: FAQs](#) tool provides guidance in such referrals.
- Consider referral for assessment of functioning:
 - if the patient has never been assessed during adolescence or adulthood.
 - if a life transition is expected (e.g., cessation of schooling or transition from middle to old age) for future planning needs (e.g., vocational or housing plans).
 - to determine contributing factors to problem behaviour or before a psychiatric diagnosis.
 - when a specific diagnosis is needed for income support services, e.g., the federal Disability Tax Credit; in Ontario, the Ontario Disability Support Program.

4. Associated Conditions and Atypical Presentations

- Appendix 2 to this Primer lists the Guidelines that focus on the health conditions and needs of adults with DD that diverge from those of the general population. The full guidelines, with references and levels of evidence, are available online in [French](#) and [English](#). In summary, the leading causes of mortality and morbidity show a different pattern of conditions than the general population [8].
 - Mortality: the top three causes are:
 - respiratory illnesses
 - cardiac disease (congenital defects as well as ischemic heart disease)
 - gastrointestinal diseases
 - Morbidity:
 - aspiration and asthma
 - dysphagia, dental problems, GERD, constipation and obesity
 - undiagnosed hearing and vision problems
 - epilepsy
 - spasticity and osteoporosis
 - hypothyroidism
 - behavioural problems
 - mental health problems
 - polypharmacy
 - physical, sexual and emotional abuse
- Sexuality is an important issue that is often not considered.
- Prevention and screening for infectious diseases and cancer are less likely to occur in this population.
- Serious illnesses can present in atypical ways, not least because of difficulties in communication.
- Behavioural change (e.g., irritability, aggression, self-injurious behaviour) may be a way of communicating physical symptoms, such as pain, or may result from the stress of environmental or social changes.
- Mental health issues are also more common in this population.
- Distinguishing the cause of behavioural change involves a biopsychosocial approach that is familiar to family physicians. [Tools](#) to help understand and assess behavioural and mental health are available: e.g., [an algorithm](#) for understanding behavioural problems and emotional concerns; [a guide](#) to prevention and management of behavioural crises; [an audit tool](#) to help ensure appropriate use of psychotropic medications.

References for Part 2: Primer for Family Physicians

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Appendix 1

Guidelines Relating to Competencies 1 – 3

Based on Sullivan et al. [Primary care of adults with developmental disabilities: Canadian consensus guidelines.](#)

General Issues	Recommendations
<p>1. Disparities in primary care exist between adults with DD and the general population. The former often have poorer health, increased morbidity, and earlier mortality [2]. Assessments that attend to the specific health issues of adults with DD can improve their primary care [9].</p>	<p>a. Apply age- and sex-specific guidelines for preventive health care as for adults in the general population [10,11]. Perform an annual comprehensive preventive care assessment including physical examination and use guidelines and tools adapted for adults with DD [9].</p>
<p>2. Etiology of DD is useful to establish, whenever possible, as it often informs preventive care or treatment [12-14].</p>	<p>a. Contact a genetics centre for referral criteria and testing protocols concerning etiologic assessment of adults whose DD is of unknown or uncertain origin [15-17].</p>
<p>Advances in genetic knowledge continue to enhance detection of etiology [13,18].</p>	<p>b. Consider reassessment periodically if a previous assessment was inconclusive, according to the criteria of the genetics centre [19].</p>
<p>3. Adaptive functioning can decline or improve in some adults with DD. A current assessment of intellectual and adaptive functioning helps to determine necessary care and supports, and establishes a baseline for future assessment [1,20,21].</p>	<p>a. Refer to a psychologist for assessment of functioning if the patient has never been assessed during adolescence or adulthood, or if a considerable life transition is expected (e.g., cessation of schooling or transition from middle to old age).</p>
	<p>b. Consider reassessment if indicated, comprehensively or in specific areas, to determine contributing factors to problem behaviour [22].</p>
<p>7. Capacity for voluntary and informed consent varies with the complexity and circumstances of decision making. The limited range of life experiences of some adults with DD, level of intellectual functioning, learned helplessness, and some mental health issues might impair</p>	<p>a. Always assess capacity for consent when proposing investigations or treatments for which consent is required [41].</p>

General Issues	Recommendations
<p>capacity to give informed or voluntary consent. An adult with DD assessed as incapable of some aspects of decision making (e.g., understanding or judging consequences) might still be able to convey, through verbal or other means, perspectives that can inform the judgment of a substitute decision maker [40].</p>	
<p>Communicating appropriately with adults with DD is necessary for assessing their capacity to consent and for seeking this consent [42].</p>	<p>b. Adapt the level and means of communicating to the patient’s level of intellectual and adaptive functioning [43].</p>
<p>Although some adults with DD might be incapable of giving consent, they might be able to contribute to decision making (e.g., understanding information, expressing perspectives, giving assent) with appropriate support from regular caregivers. Caregivers can also contribute to decision making. They may consent to or refuse treatment on behalf of an adult with DD who is assessed to be incapable of providing informed consent, if they are the most appropriate and available substitute decision makers according to the law [40].</p>	<p>c. Always consider the best interests of the adult with DD, including his or her perspective in pursuing or forgoing any health care intervention. Support whatever decision-making capacity is possible in adults with DD. Involve family or other caregivers to facilitate communication with, and understanding of, the adult with DD, but also be attentive to inappropriate taking over of decision making [42,44].</p>
<p>8. Advance care planning can often make a positive difference to the outcome of difficult life transitions and crises, and for end-of-life care [40,43,45].</p>	<p>a. Discuss advance care plans with adults with DD and their caregivers, especially to determine their preference of a substitute decision maker [41].</p> <p>b. Record advance care plans and review them annually, or sooner in the context of a health crisis, for appropriateness to the adult with DD’s present situation and for what needs to be implemented [43].</p>

General Issues	Recommendations
<p>9. Interdisciplinary health care is effective in addressing the complex needs of adults with DD. Ideally this would involve a family physician, nurse, and other health practitioners as required, with a coordinator, who might be the family physician, to ensure continuity of care [46,47].</p>	<p>a. Involve other available health professionals as needed [46]. To address complex physical, behavioural or mental health needs, consult available regional service coordination agencies or specialized interdisciplinary teams [48,49].</p>

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Appendix 2

Guidelines for Competency 4: Common Features and Atypical Presentations

Based on Sullivan et al. [Primary care of adults with developmental disabilities: Canadian consensus guidelines](#).

General Issues	Recommendations
<p>4. Pain and distress, often unrecognized [23], might present atypically in adults with DD, particularly those who have difficulty communicating. Nonspecific changes in behaviour might be the only indicator of medical illness or injury [24,25].</p>	<p>a. Be attentive to atypical physical cues of pain and distress using an assessment tool adapted for adults with DD [26,27].</p>
<p>Evaluation tools are available to assess the presence and intensity of pain in adults with DD [27-29].</p>	<p>b. Consider medical causes of changes in behaviour (e.g., urinary tract infection, dysmenorrhea, constipation, dental disease) [30].</p>
<p>Notes for Guideline 4</p> <p>The Disability Distress Assessment Tool (DisDAT) is designed to document the content and distress cues of persons with limited communication, based on the observations of carers and other persons who know the person well.</p> <p>The Wong-Baker FACES Pain Rating Tool is designed to facilitate accurate pain assessment of individuals with limited verbal expressive capacity.</p>	
<p>5. Multiple or long-term use of some medications by adults with DD can cause harm that is preventable. [31]</p>	<p>a. Review the date of initiation, indications, dosages, and effectiveness of all medications regularly (e.g., every 3 months) [32].</p> <p>b. Determine patient adherence capacity and recommend dosettes, blister-packs, and other aids if necessary.</p> <p>c. Watch for both typical and atypical signs of adverse effects [33]. Regularly monitor potentially toxic medications or interactions of medications (e.g., liver function tests or serum drug levels) at the recommended interval for each medication [34].</p>

General Issues	Recommendations
	<p>d. Ensure that patient and staff or caregivers are educated about appropriate use of medications, including over-the-counter, alternative, and as-needed medications.</p>
<p>Note for Guideline 5</p> <p>It is especially important to reassess the need for ongoing medications with this population at regular intervals, because multiple and/or long-term use of some medications (e.g., psychotropic) commonly prescribed to patients with DD may cause problems due to interactions, side effects, or atypical responses. Adults with DD may be unable to communicate these occurrences.</p>	
<p>6. Abuse and neglect of adults with DD occur frequently and are often perpetrated by people known to them [35-39]. Behavioural indicators that might signal abuse or neglect include unexplained change in weight, noncompliance, aggression, withdrawal, depression, avoidance, poor self-esteem, inappropriate attachment or sexualized behaviour, sleep or eating disorders, and substance abuse [35].</p>	<p>a. Screen annually for risk factors (e.g., caregiver stress) and possible behavioural indicators of abuse or neglect [35].</p> <p>b. When abuse or neglect is suspected, report to the police or other appropriate authority and address any consequent health issues (e.g., through appropriate counselling) [35].</p>
Physical Health Guidelines	Recommendations
<p>10. Physical inactivity and obesity are prevalent among adults with DD and are associated with adverse outcomes, including cardiovascular disease, diabetes, osteoporosis, constipation, and early mortality [50,51]. Being underweight, with its attendant health risks, is also common [52].</p>	<p>a. Monitor weight and height regularly and assess risk status using body mass index, waist circumference, or waist-hip ratio measurements [53,54].</p>
<p>A health promotion program can improve attitudes toward physical activity and satisfaction with life [55,56].</p>	<p>b. Counsel patients and their caregivers annually or more frequently, if indicated, regarding guidelines for nutrition and physical fitness and how to incorporate regular physical activity into daily routines. Refer to dietitian if indicated [56-59].</p>

Physical Health Guidelines	Recommendations
<p>Note for Guideline 10</p> <p>The Special Olympics website contains information for adults with DD and their caregivers about the benefits of physical activity, healthy lifestyles, and sport. Resources for adults with DD include:</p> <p>Strive and Train: Home Nutrition Nutrition Placemat</p> <p>Strive and Train: Home Exercise</p>	
<p>11. Vision and hearing impairments among adults with DD are often underdiagnosed and can result in substantial changes in behaviour and adaptive functioning [60-64].</p>	<p>a. Perform office-based screening of vision and hearing (e.g., Snellen eye chart, whispered voice test) annually as recommended for average-risk adults, and when symptoms or signs of visual or hearing problems are noted, including changes in behaviour and adaptive functioning [33,65].</p> <p>b. Refer for vision assessment to detect glaucoma and cataracts every 5 years after age 45 [65].</p> <p>c. Refer for hearing assessment if indicated by screening and for age-related hearing loss every 5 years after age 45 [65].</p> <p>d. Screen for and treat cerumen impaction every 6 months [66,67].</p>
<p>Notes for Guideline 11</p> <p>Eyes/Vision:</p> <p>It is important to assess the person’s ability to read and communicate to the tester the letters/symbols on the visual acuity chart.</p> <p>For patients who cannot read letters, “Tumbling E” charts or picture charts (i.e., Patti Pics) can be used. Before conducting the test, it can be helpful to have the person view the chart up close to identify each letter/image that is part of the chart.</p> <p>A card with the alphabet or images used on the chart can be given to a person who is unable or unwilling to respond verbally to allow them to identify the letters/images on the chart by pointing to the matching image on the card.</p>	

Physical Health Guidelines	Recommendations
<p>Diagnostic method:</p> <p>Ocular inspection, eye movements, visual attention, and fixation>0.2</p> <p>Visual fields (confrontation method)>0.2</p> <p>Picture Chart.....>3-4</p> <p>Tumbling E>4-5</p> <p>Snellen chart>6</p> <p>(International Organization for the Scientific Study of Intellectual Disabilities Guidelines to Visual Impairment in ID)</p>	<p>Applicable for developmental age (years):</p>
<p>Ears/Hearing:</p> <p>Screening of hearing function should be modified to account for the individual’s developmental age, delayed reaction time, behavioural problems or communication needs. Subjective audiometry in adults with developmental disabilities requires specially trained and experienced audiologists or speech and hearing therapists. However, the whispered speech test may be a useful, easy way of screening hearing when audiometry is not available or refused. It can be used for individuals who are able to repeat a series of words, whispered at a distance of three metres.</p> <p>Because of the higher incidence of excessive cerumen and cerumen impaction in adults with an intellectual disability than in the general adult population, it is important to check for occlusion of the ear canals as a possible cause of hearing loss.</p> <p>Diagnostic method:</p> <p>Behavioural observation audiometry>0</p> <p>Pure tone audiometry with visual reinforcement....>1</p> <p>Pure tone (play) audiometry>3-4</p> <p>Whispered speech at 3m>5-6</p> <p>(International Organization for the Scientific Study of Intellectual Disabilities Guidelines to Hearing Impairment in ID)</p>	<p>Applicable for developmental age (years):</p>
<p>12. Dental disease is among the most common health problems in adults with DD owing to their difficulties in maintaining oral hygiene routines and accessing dental care. Changes in behaviour can be the result of discomfort from dental disease [33,68].</p>	<p>a. Promote regular oral hygiene practices and other preventive care (e.g., fluoride application) by a dental professional [69-72].</p>

Physical Health Guidelines	Recommendations
<p>Notes for Guideline 12</p> <p>It is important to inspect the oral cavity and teeth, bearing in mind that the population of adults with DD has a higher rate of poor oral hygiene, gingivitis and periodontitis than members of the general public. Reasons for poor oral health include:</p> <ul style="list-style-type: none"> • difficulty with dental care activities (e.g., teeth brushing) • impediments to accessing a dental professional regularly • decay caused by sweetened prescription medication • altered salivary flow caused by certain medical conditions or psychotropic medication • increased incidence of bruxism in certain medical conditions (e.g., cerebral palsy) • overgrowth of gingival tissue caused by medication (e.g., Dilantin) • orofacial malformations <p>(Waldman, H.B. et al. (2001) Children with Mental Retardation/Developmental Disabilities: Do Physicians Ever Consider Needed Dental Care? Mental Retardation 39,1:53-56.)</p>	
<p>13. Cardiac disorders are prevalent among adults with DD. Risk factors for coronary artery disease include physical inactivity, obesity, smoking, and prolonged use of some psychotropic medications [51,73,74].</p>	<p>a. When any risk factor is present, screen for cardiovascular disease earlier and more regularly than in the general population and promote prevention (e.g., increasing physical activity, reducing smoking) [73].</p>
<p>Some adults with DD have congenital heart disease and are susceptible to bacterial endocarditis.</p>	<p>b. Refer to a cardiologist or adult congenital heart disease clinic [75].</p>
	<p>c. Follow guidelines for antibiotic prophylaxis for those few patients who meet revised criteria [76].</p>
<p>14. Respiratory disorders (e.g., aspiration pneumonia) are among the most common causes of death for adults with DD. Swallowing difficulties are prevalent in those patients with neuromuscular dysfunction or taking certain medications with anticholinergic side effects, and they might result in aspiration or asphyxiation [77-79].</p>	<p>a. Screen at least annually for possible signs of swallowing difficulty and overt or silent aspiration (e.g., throat clearing after swallowing, coughing, choking, drooling, long mealtimes, aversion to food, weight loss, frequent chest infections). Refer as appropriate [80].</p>

Physical Health Guidelines	Recommendations
<p>15. Gastrointestinal and feeding problems are common among adults with DD. Presenting manifestations are often different than in the general population and might include changes in behaviour or weight [81-83].</p>	<p>a. Screen annually for manifestations of GERD and manage accordingly. If introducing medications that can aggravate GERD, monitor more frequently for related symptoms [83,84].</p> <p>b. If there are unexplained gastrointestinal findings or changes in behaviour or weight, investigate for constipation, GERD, peptic ulcer disease, and pica [82,84].</p>
<p>Adults with DD might have an increased risk of Helicobacter pylori infection related to factors such as having lived in a group home, rumination, or exposure to saliva or feces due to personal behaviour or environmental contamination [83,85,86].</p>	<p>c. Screen for H pylori infection in symptomatic adults with DD or asymptomatic ones who have lived in institutions or group homes. Consider retesting at regular intervals (e.g., 3–5 years) [83].</p> <p>d. Consider urea breath testing, fecal antigen testing, or serologic testing depending on the indication, availability, and tolerability of the test [83,85].</p>
<p>Notes for Guidelines 15 and 21</p> <p>Ano-Rectum/Prostate:</p> <p>Surrey Place Centre has produced a series of eight “Health Booklets” that use clear and simple language to help prepare people with developmental disabilities for different clinical experiences, including:</p> <p>What I Need to Know about Men’s Health (Prostate/ Male Genital Exam)</p> <p>What I Need to Know about a Colonoscopy</p>	
<p>16. Sexuality is an important issue that is often not considered in the primary care of adolescents and adults with DD [87,88].</p>	<p>a. Discuss the patient’s or caregiver’s concerns about sexuality (e.g., menstruation, masturbation, fertility and genetic risks, contraception, menopause) and screen for potentially harmful sexual practices or exploitation. Offer education and counselling services adapted for those with DD [89,90].</p>

Physical Health Guidelines	Recommendations
<p>Notes for Guidelines 16 and 21</p> <p>Genitalia:</p> <p>Because of issues around communication, consent, and vulnerability to abuse, it is important to spend time with the adult patient with DD to explain the genital exam and why you are performing it. (Surrey Place “Health Booklets” can be helpful communication tools.)</p> <p>When positioning the female patient with DD on the exam table for the pelvic exam, be aware of any concerns that may cause her discomfort, such as: impaired balance/ weakness, spasticity, contractures, skin pressure over decubitus areas, inability to communicate when lying down.</p> <p>(The American College of Gynaecology (ACOG) Interactive Site for Clinicians Serving Women with Disabilities: “Part II, Module 1, The GYN exam”)</p>	
<p>17. Musculoskeletal disorders (e.g., scoliosis, contractures, and spasticity, which are possible sources of unrecognized pain) occur frequently among adults with DD and result in reduced mobility and activity, with associated adverse health outcomes [51,91].</p>	<p>a. Promote mobility and regular physical activity [56,92].</p> <p>b. Consult a physical or occupational therapist regarding adaptations (e.g., wheelchair, modified seating, splints, orthotic devices) and safety [92].</p>
<p>Osteoporosis and osteoporotic fractures are more prevalent and tend to occur earlier in adults with DD than in the general population [93]. In addition to aging and menopause, risk factors include severity of DD, low body weight, reduced mobility, increased risk of falls, smoking, hypogonadism, hyperprolactinemia, the presence of particular genetic syndromes (e.g., Down and Prader-Willi) [91,94-96], and long-term use of certain drugs (e.g., glucocorticoids, anticonvulsants, injectable long-acting progesterone in women) [34,97]. Diagnosis and management of osteoporosis related to the side effects of current treatments can be challenging in adults with DD.</p>	<p>c. Periodically assess risk of developing osteoporosis in all age groups of male and female patients with DD. Those at high risk warrant regular screening starting in early adulthood [94,96].</p> <p>d. Recommend early and adequate intake or supplementation of calcium and vitamin D unless contraindicated (e.g., in Williams syndrome) [94].</p>

Physical Health Guidelines	Recommendations
<p>Osteoarthritis is becoming more common with increasing life expectancy and weight gain, posing diagnostic and treatment difficulties [51,98].</p>	<p>e. Be aware of osteoarthritis as a possible source of pain [51].</p>
<p>Note for Guideline 17</p> <p>Consider MSK disorders (e.g., scoliosis, contractures and spasticity) as possible sources of unrecognized pain.</p> <p>Individuals with mobility impairments often find it difficult or impossible to use certain standard equipment found in clinic; for example, people who are not ambulatory cannot use standard-height examining tables. If an adjustable-height table is not readily available, assistance may be provided to help the person onto the exam table, using a safe manner to avoid injury to both the health care personnel and the patient.</p> <p>Consider modifications to certain exams, such as allowing the person to undergo the examination while remaining in the wheelchair, or assisting with dressing/undressing. (Preservice Health Training: "Examining a Person with a Mobility Impairment")</p>	
<p>18. Epilepsy is prevalent among adults with DD and increases with the severity of the DD. It is often difficult to recognize, evaluate, and control [99-101], and has a pervasive effect on the lives of affected adults and their caregivers [102].</p>	<p>a. Refer to guidelines for management of epilepsy in adults with DD [101].</p> <p>b. Review seizure medication regularly (e.g., every 3–6 months). Consider specialist consultation regarding alternative medications when seizures persist, and possible discontinuation of medications for patients who become seizure-free [101].</p> <p>c. Educate patients and caregivers about acute management of seizures and safety-related issues [103].</p>
<p>Note for Guideline 18</p> <p>Kerr, M, et. al. Consensus Guidelines into the management of epilepsy in adults with an intellectual disability, Journal of Intellectual Disability Research, 2009; 53(8): 687-94.</p>	
<p>19. Endocrine disorders (e.g., thyroid disease, diabetes, and low testosterone) can be challenging to diagnose in adults with DD [33,104-106]. Adults with DD have a higher incidence of thyroid disease compared with the general population [107].</p>	<p>a. Monitor thyroid function regularly. Consider testing for thyroid disease in patients with symptoms (including changes in behaviour and adaptive functioning) and at regular intervals (e.g., 1–5 years) in patients with elevated risk of thyroid disease (e.g., Down syndrome) [33].</p>

Physical Health Guidelines	Recommendations
	<ul style="list-style-type: none"> b. Establish a thyroid baseline and test annually for patients taking lithium or atypical or second-generation antipsychotic drugs [34].
<p>Currently there is no clear evidence of increased prevalence of diabetes in adults with DD, with some exceptions (e.g., Down syndrome) [108,109]. Diabetes management guidance has been developed for adults with DD and their care providers [110,111].</p>	<ul style="list-style-type: none"> c. Consider screening for diabetes in adults with DD who are obese or who have sedentary lifestyles or hyperlipidemia.
<p>Limited available data suggest that hypogonadism is common among men with DD [106]. Substantial data are available on hypogonadism associated with specific syndromes (e.g., Prader-Willi syndrome) [112]</p>	<ul style="list-style-type: none"> d. Consider screening for hypogonadism and testosterone level at least once after full puberty is achieved, ideally at around age 18 years, and refer as appropriate if low levels are found [105,106].
<p>20. Infectious disease prevention and screening. Even though immunization is a crucial component of preventive care, adults with DD might have limited awareness of immunizations [9,33,113].</p>	<ul style="list-style-type: none"> a. Follow guidelines for routine immunization of adults [114,115].
	<ul style="list-style-type: none"> b. Ensure influenza and Streptococcus pneumoniae vaccinations are current and offered when appropriate [116].
	<ul style="list-style-type: none"> c. Discuss the human papillomavirus vaccine with female patients with DD between the ages of 9 and 26 years and, if appropriate, their substitute decision-makers [117].
<p>It is important to screen for infectious diseases (e.g., hepatitis B, HIV, and H pylori) in adults with DD.</p>	<ul style="list-style-type: none"> d. Screen for infectious diseases based on the patient’s risk factors for exposure (for H pylori see Recommendations 15c, 15d).
<p>Some adults with DD have an increased risk of exposure to infectious diseases (e.g., hepatitis A and B) [118,119].</p>	<ul style="list-style-type: none"> e. Offer hepatitis A and B screening and immunization to all at-risk adults with DD [117-119], including those who take potentially hepatotoxic medications or who have ever lived in institutions or group homes [115].

Physical Health Guidelines	Recommendations
<p>Notes for Guidelines 20 and 21</p> <p>Consider patient’s ability to tolerate procedures (i.e., injections, mammograms) and make appropriate adjustments (i.e., anaesthetize injection site, consider sedation, US as a potential back up for mammogram). Improved communication with the patient in advance of the procedure may improve his/her ability to tolerate it. (See Surrey Place Centre’s Health Booklet Series.)</p> <p>Consider the patient’s risk factors (e.g., human papillomavirus for female patients between 9 and 26, hepatitis A and B for patients who have lived in institutions or group homes), since patients with DD may be less likely to seek out these immunizations.</p>	
<p>21. Cancer screening is an essential aspect of preventive care. However, adults with DD are less likely than those in the general population to be included in preventive screening programs such as cervical screening [113], breast examination, mammography, and digital rectal examination [2]. They are also less likely to do self-examination or to report abnormalities. Colorectal cancer risk is considerably greater for women than for men with DD [120].</p>	<ul style="list-style-type: none"> a. Perform regular cervical screening for all women who have been sexually active [121]. b. Perform annual breast screening, including mammography, for female patients with DD aged 50-69 years [122]. c. Perform an annual testicular examination for all male patients with DD [123]. d. Screen for prostate cancer annually using digital rectal examination from age 45 years for all male patients with DD [124]. e. Screen for colon cancer regularly in all adult patients with DD older than 50 years [120,125].
<p>Notes for Guidelines 15, 16, 20 and 21</p> <p>15, 21. Ano-Rectum/Prostate:</p> <p>Surrey Place Centre has produced a series of eight “Health Booklets” that use clear and simple language to help prepare people with developmental disabilities for different clinical experiences, including:</p> <p>What I Need to Know about Men’s Health (Prostate/Male Genital Exam)</p> <p>What I Need to Know about a Colonoscopy</p> <p>16, 21. Genitalia:</p> <p>Because of issues around communication, consent and vulnerability to abuse, it is important to spend time with the adult patient with DD to explain the genital exam and why you are performing it. (Surrey Place “Health Booklets” can be helpful communication tools.)</p>	

Physical Health Guidelines	Recommendations
	<p>When positioning the female patient with DD on the exam table for the pelvic exam, be aware of any concerns that may cause her discomfort, such as: impaired balance/ weakness, spasticity, contractures, skin pressure over decubitus areas, inability to communicate when lying down.</p> <p>(The American College of Gynaecology (ACOG) Interactive Site for Clinicians Serving Women with Disabilities: "Part II, Module 1, The GYN exam")</p> <p>20, 21. Infectious disease prevention and screening:</p> <p>Consider patient’s ability to tolerate procedures (i.e., injections, mammograms) and make appropriate adjustments (i.e., anaesthetize injection site, consider sedation, US as a potential back up for mammogram). Improved communication with the patient in advance of the procedure may improve his/her ability to tolerate it. (See Surrey Place Centre’s Health Booklet Series.)</p> <p>Consider the patient’s risk factors (e.g., human papillomavirus for female patients between 9 and 26, hepatitis A and B for patients who have lived in institutions or group homes), since patients with DD may be less likely to seek out these immunizations.</p> <p>21. Cancer screening:</p> <p>Surrey Place Centre has produced a series of eight “Health Booklets” that use clear and simple language to help prepare people with developmental disabilities for different clinical experiences, including:</p> <p>What I Need to Know about Breast Health</p> <p>What I Need to Know about Menopause</p> <p>What I Need to Know about a Pelvic Exam</p>

Behavioural and Mental Health Guidelines	Recommendations
<p>22. Problem behaviour, such as aggression and self-injury, is not a psychiatric disorder but might be a symptom of a health-related disorder or other circumstance (e.g., insufficient supports) [25,126,127].</p>	<p>a. Before considering a psychiatric diagnosis, assess and address sequentially possible causes of problem behaviour, including physical (e.g., infections, constipation, pain), environmental (e.g., changed residence, reduced supports), and emotional factors (e.g., stress, trauma, grief) [127].</p>
<p>Problem behaviours sometimes occur because environments do not meet the developmental needs of the adult with DD [128].</p>	<p>b. Facilitate “enabling environments” to meet these unique developmental needs, as they will likely diminish or eliminate these problem behaviours [128].</p>

Behavioural and Mental Health Guidelines	Recommendations
<p>Despite the absence of an evidence base, psychotropic medications are regularly used to manage problem behaviours among adults with DD [129,130]. Antipsychotic drugs should no longer be regarded as an acceptable routine treatment of problem behaviours in adults with DD [131].</p>	<p>c. Regularly audit the use of prescribed psychotropic medication, including those used as needed [132]. Plan for a functional analysis (typically performed by a behavioural therapist or psychologist) and interdisciplinary understanding of problem behaviours. Review with care providers psychological, behavioural, and other non-medication interventions to manage problem behaviours. Consider reducing and stopping, at least on a trial basis, medications not prescribed for a specific psychiatric diagnosis [133].</p>
<p>Notes for Guideline 22</p> <p>Risk Assessment Tool for Adults with DD in Behavioural Crisis A Guide to Understanding Behavioural Problems and Emotional Concerns Auditing Psychotropic Medication Therapy</p>	
<p>23. Psychiatric disorders and emotional disturbances are substantially more common among adults with DD, but their manifestations might mistakenly be regarded as typical for people with DD (i.e., “diagnostic overshadowing”). Consequently, coexisting mental health disturbances might not be recognized or addressed appropriately [21,134,135].</p>	<p>a. When screening for psychiatric disorder or emotional disturbance, use tools developed for adults with DD according to their functioning level (e.g., Aberrant Behaviour Checklist-Community [ABC-C]; Psychiatric Assessment Schedule for Adults with DD [PAS-ADD]) [136-139].</p>
<p>Increased risk of particular developmental, neurologic, or behavioural manifestations and emotional disturbances (i.e., “behavioural phenotypes”) is associated with some DD syndromes [140,141].</p>	<p>b. Consult available information on behavioural phenotypes in adults with DD due to specific syndromes [142,143].</p>
<p>Establishing a diagnosis of a psychiatric disorder in adults with DD is often complex and difficult, as these disorders might be masked by atypical symptoms and signs [21,135]. In general, mood,</p>	<p>c. When psychiatric disorder is suspected, seek interdisciplinary consultation from clinicians knowledgeable and experienced in DD.</p>

Behavioural and Mental Health Guidelines	Recommendations
<p>anxiety, and adjustment disorders are underdiagnosed [144] and psychotic disorders are over-diagnosed in adults with DD [145,146].</p>	
<p>24. Psychotic disorders are very difficult to diagnose when delusions and hallucinations cannot be expressed verbally [145]. Developmentally appropriate fantasies and imaginary friends might be mistaken for delusional ideation, and self-conversation for hallucination [145,147,148].</p>	<p>a. Seek interdisciplinary input from specialists in psychiatry, psychology, and speech-language pathology with expertise in DD to help clarify diagnoses in patients with limited or unusual use of language [144,149,150].</p>
<p>25. Input and assistance from adults with DD and their caregivers are vital for a shared understanding of the basis of problem behaviours, emotional disturbances and psychiatric disorders, and for effectively developing and implementing treatment and interventions [127,151,152].</p>	<p>a. Establish a shared way of working with patients and caregivers. Seek input, agreement, and assistance in identifying target symptoms and behaviours that can be monitored.</p>
	<p>b. Use tools (e.g., sleep charts, antecedent-behaviour-consequence [ABC] charts) to aid in assessing and monitoring behaviour and intervention outcomes [153,154].</p>
<p>26. Interventions other than medication are usually effective for preventing or alleviating problem behaviours [133,144,155].</p>	<p>a. To reduce stress and anxiety that can underlie some problem behaviours, emotional disturbances, and psychiatric disorders, consider such interventions as addressing sensory issues (e.g., under-arousal, over-arousal, hypersensitivity), environmental modification, education and skill development, communication aids, psychological and behaviour therapies, and caregiver support [144].</p>
	<p>b. Cognitive behavioural therapy can be effective in decreasing anger and treating anxiety and depression in adults with DD [156,157].</p>

Behavioural and Mental Health Guidelines	Recommendations
	<p>c. There is increasing evidence of the efficacy of psychotherapy for emotional problems (e.g., related to grief, abuse, trauma) that might underlie aggression, anxiety, and other such states [158-162].</p>
<p>27. Psychotropic medications (e.g., antidepressants) are effective for robust diagnoses of psychiatric disorders in adults with DD[163] as in the general population [164].</p>	<p>a. When psychiatric diagnosis is confirmed after comprehensive assessment, consider psychotropic medication along with other appropriate interventions as outlined in guideline 26 [165].</p>
<p>Psychotropic medications, however, can be problematic for adults with DD and should therefore be used judiciously. Patients might be taking multiple medications and can thus be at increased risk of adverse medication interactions. Some adults with DD might have atypical responses or side effects at low doses. Some cannot describe harmful or distressing effects of the medications that they are taking [34,166].</p>	<p>b. “Start low, go slow” in initiating, increasing, or decreasing doses of medications [167].</p> <p>c. Arrange to receive regular reports from patients and their caregivers during medication trials, in order to monitor safety, side effects, and effectiveness [133].</p> <p>d. In addition to reviews every 3 months (see guideline 5), also review the psychiatric diagnosis and the appropriateness of prescribed medications for this diagnosis whenever there is a behavioural change [34,133].</p>
<p>When unable to pinpoint a specific psychiatric diagnosis, behaviours of concern might serve as index behaviours against which to conduct a trial of medications [133,167].</p>	<p>e. Having excluded physical, emotional, and environmental contributors to the behaviours of concern, a trial of medication appropriate to the patient’s symptoms might be considered.</p>
<p>28. Antipsychotic medications are often inappropriately prescribed for adults with behaviour problems and DD [168]. In the absence of a robust diagnosis of psychotic illness, antipsychotic medications should not be regarded as routine treatments of problem behaviours in adults with DD [131].</p>	<p>a. Do not use antipsychotic medication as a first-line treatment of problem behaviours without a confirmed robust diagnosis of schizophrenia or other psychotic disorder [131].</p>

Behavioural and Mental Health Guidelines	Recommendations
<p>Antipsychotic medications increase risk of metabolic syndrome and can have other serious side effects (e.g., akathisia, cardiac conduction problems, swallowing difficulties, bowel dysfunction) [34,166].</p>	<p>b. Carefully monitor for side effects of antipsychotic medication, including metabolic syndrome. Educate patients and caregivers to incorporate a healthy diet and regular exercise into their lifestyle [34].</p> <p>c. Reassess the need for ongoing antipsychotic medications at regular intervals and consider dose reduction or discontinuation when appropriate (also see guidelines 5 and 27) [34].</p>
<p>29. Behavioural crises can occasionally arise that might need management in an emergency department [169-173].</p>	<p>a. When psychotropic medications are used to ensure safety during a behavioural crisis, ideally such use should be temporary (no longer than 72 hours).</p> <p>b. Debrief with care providers in order to minimize the likelihood of recurrence. This should include a review of crisis events and responses (e.g., medication, de-escalation measures), and identification of the possible triggers and underlying causes of the behavioural crisis [133,174].</p> <p>c. If the patient is at risk of recurrent behavioural crises, involve key stakeholders, including local emergency department staff, to develop a proactive, integrated emergency response plan [174].</p>
<p>30. Alcohol or drug abuse is less common among adults with DD than in the general population, but the former might have more difficulty moderating their intake and experience more barriers to specialized rehabilitation services [175-177].</p>	<p>a. Screen for alcohol and drug abuse as part of the annual health examination.</p>

Behavioural and Mental Health Guidelines	Recommendations
<p>31. Dementia is important to diagnose early, especially in adults with Down syndrome who are at increased risk [178]. Diagnosis might be missed because changes in emotion, social behaviour, or motivation can be gradual and subtle. A baseline of functioning against which to measure changes is needed.</p>	<p>a. For patients at risk of dementia, assess or refer for psychological testing to establish a baseline of cognitive, adaptive, and communicative functioning. Monitor with appropriate tools [179].</p>
<p>Differentiating dementia from depression and delirium can be especially challenging [180].</p>	<p>b. Educate family and other care providers about early signs of dementia. When signs are present, investigate for potential reversible causes of dementia.</p>
	<p>c. Consider referral to the appropriate specialist (i.e., psychiatrist, neurologist) if it is unclear whether symptoms and behaviour are due to emotional disturbance, psychiatric disorder, or dementia [179].</p>
<p>Note for Guideline 31</p> <p>The Assessments for Adults With Developmental Disabilities is a tool to be filled out with caregivers of adult patients with DD with suspected dementia.</p>	

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Family Medicine Curriculum Resource: Adults with Developmental Disabilities

Part 3: Case Modules



Family Medicine Curriculum Resource: Adults with Developmental Disabilities

Part 3 – Case Modules*

Introduction

The Primary care of adults with developmental disabilities: Canadian consensus guidelines ([English](#) and [French](#)) were initially published in 2006 and were updated in 2011. They are accompanied by several Tools that have been designed to facilitate their implementation. The Guidelines are divided into three sections: General Medical Issues, Physical Health Guidelines, and Behaviour and Mental Health Guidelines.

The following three modules contain long clinical scenarios that aim to draw the learner's attention to the Guidelines and Tools from the Primary care of adults with developmental disabilities: Canadian consensus guidelines.

The cases are divided into the same three sections as the Guidelines:

1. General Medical Issues
2. Physical Health Guidelines
3. Behaviour and Mental Health Guidelines

Their intended use is either for a facilitator to present to a group of learners to go through in a Problem-Based Learning format, or for individual learners to work through individually to familiarize themselves with the Tools and Guidelines. As the learners work through the case, they will be guided by questions that will highlight different Tools and Guidelines.

The Information Points section will contain the specific guidelines that are outlined in the case, and the Commentary section will contain a discussion of the Guidelines and Tools and highlight any other important points that are brought up in the case.

Module 1: General Medical Issues in Primary Care

Outcomes

After completing this module, the learner shall be able to:

1. Apply the General Section of the Guidelines and Tools when initiating care for a patient with DD.
2. Identify strategies to establish effective working relationships with patient and caregivers.

Overview of Tools for General Issues in Primary Care

Genetic Assessment: Frequently Asked Questions

This [document](#) provides answers to commonly asked questions about investigation of genetic etiology for DD. It includes information on why genetic assessment can be helpful and which patients should be considered for referral.

Adaptive Functioning and Communication Associated with Different Levels of Developmental Disabilities

This [Tool](#) shows the types of life skills and communication skills that usually correspond with various levels of intellectual ability.

Psychological Assessment: Frequently Asked Questions

This [document](#) provides answers to commonly asked questions about psychological assessment, including information on resources that may be considered if assessment is not covered by a private medical plan, and how psychological assessment can benefit a person with DD.

Informed Consent in Adults with Developmental Disabilities

This [Tool](#) discusses steps involved in the consent process, provides sample questions for a capacity assessment, outlines the hierarchy for identification of a substitute decision-maker in Ontario, and makes recommendations on documentation. It also outlines the consent required for incapable patients in various medical situations in Ontario, and provides a list of federal and provincial informed consent legislation websites.

Communicating Effectively with People with Developmental Disabilities

This [Tool](#) provides communication tips for establishing rapport, choosing appropriate language, listening, explaining clearly, and communicating without words. It also lists communication-related resources.

Office Organizational Tips – Patients with Developmental Disabilities

This [Tool](#) gives organizational tips for pre-appointment preparation, the initial office appointment, follow-up visits, exams and investigations, referrals, and prescriptions. It also lists further resources.

Today's Visit

This [Tool](#) is a form that can be updated by the caregiver prior to each doctor's appointment. It serves to update the physician and/or nurse, and helps to ensure patient and caregiver concerns are addressed and documented. It also includes a monitoring tool for activity level, sleep, eating/weight, bowel routine, and mood/behaviour.

Advocacy Role of Family Physician/Advanced Practice Nurse

This [document](#) outlines the advocacy role of the physician/nurse in caring for an individual with DD. Topics of importance include patient-centred care, ensuring optimal quality of life and appropriate caregiver expectations, communicating service gaps, and being knowledgeable about regulations, policies, and initiatives.

Community Resources in Ontario

This [document](#) provides information about the Ministry of Community and Social Services, Community Networks of Specialized Care, ConnexOntario, Respite Services, and the Centre for Addiction and Mental Health.

Sally, female, age 55

Part 1:

Sally is a woman with severe developmental disability (DD) and cerebral palsy who is at your office today for her first visit. A caregiver from her new group home accompanies her. The discharge summary from the regional centre where she had lived for the past 50 years gives the following information:

Past medical history

- DD of unknown etiology; genetic assessment at age 28 was inconclusive
- IQ 25 - 40; psychological assessment at age 6
- Cerebral palsy with spastic quadriplegia and dysarthric speech
- Seizure disorder; last seizure was two years ago
- Dysphagia and history of recurrent aspiration pneumonia; G-tube feedings
- Gastroesophageal reflux disease
- Chronic constipation
- Post-menopausal
- Osteopenic spine
- Eczema

Past surgical history

- Several tendon releases for flexion contractures
- Paraspinal rod insertion for scoliosis
- G-tube insertion

Medications

- Ferrous sulfate, 2 mL BID
- Lactulose, 40 mL, q 2 days
- Clonazepam, 0.5 mg, 2 tabs q am; 1 tab at noon; 2 tabs qhs
- Betamethasone cream, prn

Social history

- Her Substitute Decision-Maker is her younger sister
- Dependent on caregivers for activities of daily living and requires wheelchair for mobility
- No formal education or employment
- Single; no history of sexual relationship
- No smoking or alcohol use

Family history

- Parents deceased
- Twin died in utero at 36 weeks of GA
- Four siblings, three older and one younger; none with DD

Given what you know about Sally, do you expect that she will be able to participate in the discussion about her care?

Will she be able to participate in decisions about her medical care?

Part 2:

When Sally comes into the office with her caregiver, you introduce yourself and ask Sally how she's doing at her new home. Her dysarthric speech and limited vocabulary make her response difficult to understand. You explain to Sally that you'd like to ask her caregiver some questions about her, and ask if that's okay. Sally nods.

Sally's caregiver tells you the G-tube feedings have been implemented without difficulty, but personal care has been awkward, as the approaches differ from those Sally is used to.

She has noticed that Sally's clothes hang loosely and she is concerned Sally may be losing weight. They are unable to weigh her at the group home, but they know that five years ago her BMI was 20 and suspect it is now lower.

On further questioning you learn that Sally has been fed with a G-tube since she was 17 and had her last formal nutritional assessment at that time. She receives Peptamen 1.5 one can TID and 800 mL H₂O daily. Sometimes she gags and vomits with feeding, and volume reductions are necessary. She also has gastroesophageal reflux that has not responded to several trials of postural feeding manoeuvres.

She has a history of chronic constipation but her bowels have moved daily since she started taking Lactulose and they are now sometimes loose. She occasionally has fecal incontinence when her stools are loose, and her caregiver wonders if this may be the cause of a rash on her perineum. She asks you if you could take a look.

How can you optimize communication with Sally?

What adaptations, if any, will you make to your usual approach to doing a physical exam with Sally?

Part 3:

Sally is in a wheelchair, and you do not have the equipment available to weigh her in your office. Sally's caregiver helps you transfer Sally to the examination table.

Physical examination

- Appears thin but well nourished
- Blood pressure is 95/65 mm Hg
- Pulse rate is 68 and regular
- Waist circumference is 80 cm
- The G-tube insertion site is clean and not inflamed
- Abdomen is soft; liver, kidneys, and spleen are not palpable; there are no abdominal masses

Sally does not display any signs of discomfort during the exam. You ask Sally if you could examine the rash on her bottom and she nods. You explain that her pants will need to be removed for the exam. As her caregiver helps you with this, Sally makes a vocalization and grabs her caregiver's wrists. You notice tears in Sally's eyes.

What is your response to Sally's distress?

Part 4:

You apologize to Sally, and stop the exam. Sally is systemically well and the rash is localized and not bothersome, so you ask her caregiver to keep the area clean and dry and you defer the exam.

You decide to have Sally back in a week or two after you discuss the need for a physical exam with her sister, who is also her Substitute Decision-Maker. Sally's caregiver

suggests that Sally might be more comfortable disrobing while you are outside the room, and says she will bring a familiar blanket next time for her to cover up with until it is time for the exam.

She also reminds you that they need to have Sally's prescriptions renewed before they leave today.

What, if any, changes would you make to Sally's current medication regime?

What is your long-term plan for Sally?

Module 1 Commentary

Part 1:

- Given what you know about Sally, do you expect that she will be able to participate in the discussion?*

Understanding an individual's intellectual ability (i.e., intelligence quotient and/or age equivalence) and level of adaptive functioning (i.e., the skills a person has to handle common demands of daily living) helps the primary care practitioner to optimize clinical encounters by enabling optimal communication. ([Adaptive Functioning and Communication associated with Different Levels of Developmental Disabilities](#))

Based on the information in the Adaptive Functioning and Communication Associated with Different Levels of Developmental Disabilities tool, individuals like Sally with severe DD (IQ: 25-23 ± 5) have an age equivalence of 3-6 years. They have limited vocabulary, and may use single- and 2-word combinations, gestures and signs to communicate. Comprehension is usually limited to the immediate environment, but some action words may be understood ([Adaptive Functioning and Communication associated with Different Levels of Developmental Disabilities](#)). In Sally's case, she will likely be able understand some of the conversation, and may be able to respond to simple questions and directions like "Point to where it hurts". She may require time to process questions and formulate a response.

One caveat in this case is that Sally's most recent psychological assessment was done at age 6, and we do not know if this accurately reflects her current abilities because adaptive function can decline or improve in some people with DD ([Consensus Guidelines #3](#)). Referral to a psychologist for assessment would be appropriate in her case ([Consensus Guidelines #3](#)).

For more information on referring individuals with DD for Psychological Assessment, see the [Psychological Assessment: Frequently Asked Questions](#) tool.

- Will she be able to participate in decisions about her medical care?*

"Capacity for voluntary and informed consent varies with the complexity and circumstances of decision making" ([Consensus Guidelines #7](#)) and therefore needs to be assessed on a case-by-case basis. In Sally's case, it is unlikely she would be able to provide informed consent for most medical decisions because she has severe DD and would likely not have the level of intellectual function required to think abstractly and thus weigh possible outcomes of a decision. She may be able to contribute to decision-making however, and it would be important to support by involving family and caregivers in facilitating communication of her perspective ([Consensus Guidelines #7](#)).

When a patient has a mild or moderate developmental disability, it is more likely that

they may have the capacity to consent to a proposed investigation or treatment. Sample questions for a capacity assessment are found in section C of the [Informed Consent in Adults with Developmental Disabilities](#) Tool. If a patient is found incapable of consenting, a substitute decision-maker (SDM) will need to be consulted ([Consensus Guidelines #7](#)).

Part 2

□ *How can you optimize communication with Sally?*

Individuals with DD often have communication difficulties. In Sally's case, it is important to remember that her receptive language may be better than her expressive language, especially given her dysarthric speech. It is good practice to assume that a person with DD can understand more than they can communicate. A speech language pathologist can assess an individual's receptive and expressive language skills and provide specific recommendations on how to optimize communication.

A number of general communication tips are provided in the [Communicating Effectively with People with Developmental Disabilities](#) tool.

Tips include:

- speaking directly with the individual with DD
- using concrete language and simple sentences
- avoiding shouting
- listening carefully
- allowing enough time
- explaining what will happen before you begin
- being prepared to communicate with visual aids or vignettes

Setting the stage for optimal communication is important and the Surrey Place Centre [Health Booklet Series](#) aims to offer people with DD clear and simple guidance on a variety of topics.

Health-related pamphlets and videos designed for individuals with DD can also be found at www.easyhealth.org.uk.

□ *What adaptations, if any, will you make to your usual approach to doing a physical exam with Sally?*

It is always important to ask permission before encroaching on another individual's personal space. In Sally's case, it is important to gauge whether she understands what the physical exam entails, and whether she feels comfortable with it. Her primary caregiver may have some insight on how much she comprehends, and how the exam should be approached.

Some patients require ongoing reassurance and communication at each step of the exam, and some require visual aids, social stories, or desensitization techniques. For some individuals it may take several visits before they feel comfortable with any type of exam.

Positive re-enforcement, words of encouragement and distraction may be useful, but at the same time, it is important to keep in mind that it may be necessary to adjust, delay, or discontinue the exam if a patient does not assent.

It is also important to remember that pain and distress may present atypically in patients with DD, especially when those individuals have difficulty communicating, and it is important to be attentive to physical cues ([Consensus Guidelines #4](#)).

In Sally's case, adjustments to the exam would also be necessary due to her physical disability. Extra time and flexibility with examination techniques may be required (e.g., measuring waist circumference instead of weighing).

More tips on performing physical exams and investigations can be found in the [Office Organizational Tips](#) Tool.

Part 3

❑ *What is your response to Sally's distress?*

It is important to acknowledge Sally's distress, and to stop the exam until consent and assent have been obtained. Given Sally's level of DD and communication limitations, it is likely her SDM would need to be consulted prior to invasive exams, procedures and treatments. In Sally's case, her SDM may have provided consent for physical examination and common investigations by agreeing to "plan of care" at her group home. If this were the case, it would still be necessary to seek assent from Sally before proceeding.

This case serves as a reminder that people with DD are members of a vulnerable population and they are unfortunately at high risk of abuse. The consensus guidelines recommend screening for signs of abuse and neglect at least annually ([Consensus Guidelines #6](#)).

Part 4:

❑ *What, if any, changes would you make to Sally's current medication regime?*

Time with Sally is limited at this visit and there are gaps in her medical history, so it may not be possible to do a full medication review. Review of medications should be a priority in future visits, however. The consensus guidelines point out that individuals with DD may be prescribed multiple medications for comorbid conditions, including behavioural and mental health problems, and they may not be able to communicate side

effects. It is recommended that indications, efficacy, and dosage of medications be reviewed regularly (e.g., every three months) ([Consensus Guidelines #5](#)).

Interdisciplinary coordination of care is often required for individuals with DD ([Consensus Guidelines #9](#)) and, in Sally's case, the expertise of a pharmacist could be drawn upon (especially around issues of timing and route of medication administration with enteral feeding). A dietician could also provide valuable information on ensuring Sally receives adequate caloric, fluid, fibre, vitamin and mineral intake.

❑ *What is your long-term plan for Sally?*

Sally's weight loss will need to be investigated, and it will be important to address consent issues with Sally's SDM, and obtain Sally's assent for these investigations. It will also be important to anticipate future health care needs (e.g., planning for health-related crises) with Sally's SDM and caregivers ([Consensus Guidelines #8](#)).

It will likely take several visits to obtain Sally's history, to perform a full physical exam, and to address preventive health care issues. In Sally's case, the *Cerebral Palsy Health Watch Table* (available soon on the Surrey Place website) would be of particular use. The recommendations are organized by system and are specific to individuals with cerebral palsy.

In order to optimize Sally's care, it would be ideal to determine the etiology of her developmental disability. Guidelines recommend a comprehensive etiological assessment including genetic testing with periodic reassessment ([Consensus Guidelines #2](#)). More information on Genetic testing can be found in the [Genetic Assessment: Frequently Asked Questions](#) Tool.

Health care needs of individuals with DD are often complex. Primary care providers often act as care coordinators and advocate for their patients to receive equitable, person-centred care. Instructions on how to access information on laws, regulations, policies, initiatives and services relevant to persons with DD can be found in the [Advocacy Role of Family Physician/Advanced Practice Nurse](#) Tool.

References for Module 1 of Part 3: Case Modules

1. Sullivan et al. Primary care of adults with developmental disabilities. Canadian Consensus Guidelines. *Can Fam Physician* 2011;57: 541-53.
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Module 2: Physical Health Guidelines

This module will focus on the second section “Physical Health Guidelines” and the corresponding Tools.

Outcomes

By the end of the module, the learner will be able to:

1. Apply the Physical Health Guidelines and Tools to a case study of a patient with a developmental disability
2. Utilize the Cumulative Patient Profile tool adapted for patients with developmental disabilities
3. Utilize a Health Watch Table for care of a patient with a syndrome-specific developmental disability

Overview of Physical Health Tools

Cumulative Patient Profile for adults with developmental disabilities

This [Tool](#) may be used by the primary care practitioner to keep an ongoing, up-to-date record of the patient’s current problem list, past medical history, current and past medications, allergies, immunizations, family history, social history, personal history, and special needs and methods of communications for use to prepare for appointments and to record changes during appointments.

Preventive Care Checklist Form for adult females and Preventive Care Checklist Form for adult males

These Tools, which are specific for [male](#) and [female](#) patients, are preventive care checklist forms that can be used at a periodic health exam for patients with a developmental disability for which there is no known etiology, or no syndrome-specific Health Watch Table. Based on the College of Family Physicians of Canada’s endorsed Preventive Care Checklist for the general population, the highlighted sections refer to specific guidelines that differ from the recommendations for the general population. These are explained in the *Explanations for Preventive Care Checklist Form – Adaptations for Adults with Developmental Disabilities* section that follows.

Health Watch Tables for Selected Developmental and Related Disabilities

These syndrome-specific Tools highlight identified health concerns that should be monitored or screened for specific etiologies of developmental disabilities.

Currently, Health Watch Tables have been developed for:

- [Down syndrome](#)
- [Fragile X syndrome](#)

- [Prader-Willi syndrome](#)
- [Smith-Magenis syndrome](#)
- [22q11.2 Deletion syndrome](#)
- [Fetal alcohol spectrum disorder](#)
- [Williams syndrome](#)

Health Watch Tables for cerebral palsy and autism spectrum disorder will be available soon on the Surrey Place website.

Emilio, male, age 46

Part 1:

Emilio is a 46-year-old man who has Down syndrome and an intellectual disability in the moderate range. He is new to your practice and has been brought in today for an initial visit by his sister, Marina, and his group home worker, Sheldon. His main health problems are obesity and hypothyroidism.

You begin by reviewing Emilio's history. Emilio came to Canada with his family from Brazil when he was 6 years old and speaks some English, but primarily Portuguese. He has three older sisters who are in good health and who have families of their own. Emilio's father died 14 years ago and, since that time, Emilio has lived with his mother in the family home. Six months ago, Emilio's mother's health deteriorated, and she became unable to care for Emilio by herself. He was moved to a group home two months later, but continues to spend most weekends at his mother's home.

Emilio has no known drug allergies. His only medication is levothyroxine 0.15 mg once daily.

He does not smoke or drink alcohol. He played golf and soccer with his father, but since his father's death, he has led a sedentary lifestyle.

During Emilio's visit, he is pleasant and cooperative. He responds to most questions with one- or two-word answers.

You conclude the visit by asking Emilio to return for a complete physical examination. You also request a copy of his medical chart from his previous family physician's office.

What are your priorities for the care of your new patient, Emilio?

What would you include in a plan for Emilio's health care going forward?

Part 2:

Emilio returns four weeks later, accompanied by his mother. You had a chance to review his old chart and found that he visited his previous family physician once or twice a year. His last periodic health exam was seven years ago, and blood work was last drawn two years ago. Emilio had a genetic assessment when he was a newborn. His last psychological assessment to assess his level of functioning was done for primary school placement when he was 10 years old and showed a moderate level of intellectual disability.

Vision, hearing, and dental assessments

Emilio wears glasses. An optometrist assesses his vision every two years. His last hearing assessment was done three years ago and was normal. He is uncooperative with regular dental work and any dental exams or treatments are done under general anaesthesia. His last dental assessment was three years ago.

Immunizations

Emilio's tetanus toxoid, pertussis and hepatitis B immunizations are up to date.

Medications

Emilio has been taking the same dose of his current medication for the past three years.

Laboratory tests (*completed two years ago*)

Complete blood count, creatinine, blood urea nitrogen, electrolytes, blood urea nitrogen, fasting blood glucose, fasting lipid profile, and thyroid-stimulating hormone (TSH).

The results were within normal limits except for an elevated TSH level of 10.0.

Physical examination

- Weight 124 kg; height 164 cm; waist circumference 132 cm; BMI 46
- Blood pressure 106/80 mmHg; heart rate 80 bpm and regular
- Cerumen impacted in both ear canals
- Visual acuity: OD 20/40, OS 20/30 (corrected)
- Periodontal disease
- Thyroid, chest, cardiovascular, and abdominal exams normal
- Genital, musculoskeletal, and neurological exams normal

- ❑ *What are Emilio's main physical health issues? How would you monitor them?*
- ❑ *What other issues regarding Emilio's health were not addressed with the past medical history review and physical exam?*

Part 3:

Nine months later, Emilio returns to the clinic with Sheldon, who is concerned about some changes in Emilio's behaviour. Emilio has become withdrawn over the past two months. Group home staff members have had trouble engaging him in activities that he used to enjoy, and he spends the day at his day program resting on a couch. He is eating more than usual and sleeping less at night.

Upon further questioning, you discover that Emilio's mother's health has deteriorated and that she moved to a seniors' complex in the city two months ago. Emilio no longer goes to visit her on weekends. Furthermore, Sheldon explains that some changes have occurred in the group home recently. A new resident who moved in six weeks ago is creating a disturbance, with loud screaming and banging on the walls at night. Staff members are struggling to control this situation.

- ❑ *What are the core issues that need to be addressed in a management plan for Emilio?*

Module 2 Commentary

Part 1:

❑ *What are your priorities for the care of your new patient, Emilio?*

When meeting any new patient, it is important to begin to develop rapport early on. This is especially important in patients with a developmental disability, as they may have heightened anxiety about attending a physician's office or meeting someone new. Take the time early on to introduce the patient to office staff, and allow them to gain comfort in the office environment. Some suggestions can be found in the [Office Organizational Tips](#) tool.

Emilio's records from his previous family physician should be reviewed to see when a periodic health exam and investigations were last done. A thorough medication review should be done to determine the date of initiation, indication, dosage and effectiveness of each medication.

The consensus guidelines recommend doing a medication review every three months to prevent the harm that can be caused by multiple or long-term use of some medications ([Consensus guidelines #5](#)). Emilio's acute medical issues should be reviewed, including his hypothyroidism and his weight.

❑ *What would you include in a plan for Emilio's health care going forward?*

Once Emilio's acute medical issues are addressed, time should be taken to address preventive care and monitoring. His current and past medical history should be reviewed and documented. The [Cumulative Patient Profile for adults with developmental disabilities \(CPP\)](#) tool may be helpful for these records and can continue to be updated with changes to the patient's health.

Once Emilio is able to come back for his periodic health exam, the [Preventive Care Checklist Form for adult males with a developmental disability \(PCC\)](#) tool can be used to help the physician focus on preventive health issues especially relevant to patients with developmental disabilities. For example, a gastrointestinal history should be taken to monitor for *Helicobacter pylori* infection, gastroesophageal reflux disease and constipation.

Given the disparities that exist in primary care between adults with developmental disabilities and the general population, the consensus guidelines recommend an annual comprehensive preventive care assessment using the guidelines and tools adapted for adults with developmental disabilities ([Consensus guidelines #1](#)).

In addition to the CPP and PCC, Health Watch Tables have been developed for certain specific syndromes with an established etiology and identified health concerns. Using the [Down Syndrome Health Watch Table](#) can help lead the physician in syndrome-

specific anticipatory care for patients with Down syndrome to determine what screening and monitoring is needed. For example, Emilio should be screened for obstructive sleep apnea, since 50 to 80% of adults with Down syndrome are affected.

The physician will need to see Emilio more frequently initially while they are establishing rapport, collecting past medical information, and establishing plans for monitoring, preventive care, and medication titration. It may also be easier for Emilio to have several short visits, instead of longer visits that might be overwhelming for him. Advice should be sought from his caregiver to help determine the best frequency for his visits. Scheduling him as the first patient of the day may be helpful if the waiting room is a difficult environment for him, or if waiting is problematic.

Emilio will need blood work done to monitor his thyroid as well as lipid and blood glucose screening given his obesity.

Part 2:

❑ *What are Emilio's main physical health issues? How would you monitor them?*

Vision and Hearing

Emilio's ears should be cleared of cerumen using mineral oil for several days, followed by irrigation. This should be monitored every six months. Emilio's hearing should be screened annually by his family physician, and hearing assessment should be done at least every two years.

His vision should be checked every one to two years, looking specifically for cataracts and significant refractive errors, which are common in people with Down syndrome ([Consensus guidelines #11](#); [Down Syndrome Health Watch Table](#)).

Dental Health

Emilio should be referred to a dentist to manage his periodontal disease and for regular clinical exams every six months. Emilio's dentist should be someone who has the ability to assess and treat Emilio under a general anesthetic ([Consensus guidelines #12](#)).

Immunizations

Discuss the possibility of *Streptococcus pneumoniae* vaccination with Emilio and his mother, and encourage yearly influenza vaccine. Adults with developmental disabilities have an increased risk of exposure to infectious diseases such as Hepatitis A and B, so Emilio should be offered Hepatitis A vaccination as well ([Consensus guidelines #20](#)).

Thyroid

Emilio's thyroid function should be tested now, given his previous elevated TSH, and then at regular intervals once stabilized. Inquire about any symptoms that could be attributed to thyroid dysfunction, including changes in sleep, behaviour, weight or adaptive function ([Consensus guidelines #19](#)). Emilio's thyroid dysfunction could also be contributing to his increased weight and low energy level.

Weight

Given Emilio's obesity, his weight, height and waist circumference or waist-to-hip ratio should be measured and his BMI calculated.

Physical inactivity and obesity are prevalent among adults with developmental disabilities and are associated with cardiovascular disease, diabetes, osteoporosis, constipation and early mortality ([Consensus guidelines #10](#)).

Emilio should be screened for cardiovascular disease regularly, as his risk is increased ([Consensus guidelines #13](#)).

Emilio's diet should be reviewed and referral to a dietician could be considered. It would also be beneficial to review what physical activity he enjoys and to establish a realistic weight loss program.

❑ *What other issues regarding Emilio's health were not addressed with the past medical history review and physical exam?*

The [Down Syndrome Health Watch Table](#) outlines several syndrome-specific considerations. Obstructive sleep apnea is more common in patients with Down syndrome and, therefore, a careful history of snoring, apnea, morning headaches and daytime sleepiness should be gathered. He should be sent for a sleep study if there are any concerns.

Fifty per cent of adults with Down syndrome have cardiovascular concerns, including acquired mitral valve prolapse and valve regurgitation. An annual cardiac exam and baseline echocardiogram can confirm any abnormalities.

Adults with Down syndrome are also at continued risk of spinal cord compression secondary to atlantoaxial instability. Annual neurological exams and a baseline lateral cervical spine x-ray should be done and repeated if there are any signs or symptoms of neurologic compromise ([Down Syndrome Health Watch Table](#)).

Part 3:

❑ *What are the core issues that need to be addressed in a management plan for Emilio?*

Emilio should continue to receive ongoing medical care, including physical exams and investigations for monitoring, screening, and to rule out any medical conditions that could be underlying his change in mood ([Consensus guidelines #4](#)). Emilio's mood should be assessed and treated if a mood disorder is diagnosed ([Consensus guidelines #23](#)), considering both medication and counselling with a professional who has experience with patients with developmental disabilities.

To further support his mood, Emilio needs to be encouraged to exercise regularly and participate in his day program and regular visits with his family. Emilio should be provided with support with the transitions he is facing, as should his family. Significant changes in the lives and health of both their mother and their brother will be extremely stressful for Emilio's sisters.

Screening for dementia should also be initiated, given his change in behaviour, as 11% of adults with Down syndrome aged 40 to 49 show signs of dementia (*Visser et al., 1997*) (3). This should be done through referral for neuropsychological testing to establish a baseline of cognitive, adaptive and communicative functioning, which can subsequently be monitored with specific tools ([Consensus guidelines #31](#)).

Periodic team meetings are a good way to engage all members of the team in reviewing the long-term treatment plan and the medications.

References for Module 2 of Part 3: Case Modules

1. Sullivan et al. [Primary Care of adults with developmental disabilities: Canadian Consensus Guidelines](#). *Can Fam Physician* 2011;57: 541-53.
2. Developmental Disabilities Primary Care Initiative. Tools for the primary care of people with developmental disabilities. 1st ed. Toronto: MUMS Guideline Clearinghouse; 2011.
3. Visser FE, Aldenkamp AP, Van Huffelen AC, et al. *Prospective study of the prevalence of Alzheimer-type dementia in institutionalized individuals with Down syndrome*. *American Journal on Mental Retardation*, 1977, 101:404-412.

Module 3: Behavioural and Mental Health Guidelines

Outcomes

After completing this module, the primary care practitioner shall be able to:

1. Utilize the Behavioural and Mental Health Guidelines and the Corresponding Tools to develop a comprehensive approach to a situation of a complex behavioural problem.
2. Consider the risks and benefits of prescribing psychotropic medication for individuals with DD.

Overview of Behavioural and Mental Health Tools

Initial Management of Behavioural Crises in Family Medicine

This [Tool](#) guides development of an initial management plan for behavioural crisis, and includes an algorithm to clarify the cause of the behaviour. It also provides recommendations on post-behavioural crisis management.

Risk Assessment Tool for Adults with DD in Behavioural Crisis

This [Tool](#) guides assessment of suicide risk, risk of self-harm and self-neglect, risk of victimization or exploitation, risk to others, and risk to environment.

A Guide to Understanding Behavioural Problems and Emotional Concerns

This [two-part Tool](#) helps to identify causes of behavioural problems in order to guide management. Part A is intended for usage by the primary care provider/team and Part B is intended for the caregiver.

ABC (Antecedent-Behaviour-Consequence) Chart

This [Tool](#) provides a template to record incongruent, challenging, or problematic behaviours along with possible triggers and consequences.

Crisis Prevention and Management Plan

This [Tool](#) provides a template for development of a crisis prevention and management plan, along with instructions on using the template and an example of a completed plan.

Essential Information for Emergency Department

This [form](#) can be filled out with pertinent information prior to an emergency department visit. It is meant to give the emergency department physician a summary of contact information, the reason for the ED visit, and an overview of health status.

Guidance about Emergencies for Caregivers

This [Tool](#) provides caregivers with tips on advocacy, communication, and safety in the emergency department during a behavioural crisis.

Psychotropic Medication Issues

This [document](#) reviews general prescribing recommendations and monitoring of psychotropic medications for adults with DD.

Auditing Medication Therapy

This [checklist](#) can be used to evaluate the appropriateness of continuing or discontinuing previously prescribed psychotropic medications.

Rapid Tranquilization of Adults with Crisis Behaviours

This [Tool](#) is meant to aid primary care providers in prescribing medications for rapid tranquilization when patients with DD are exhibiting crisis behaviours.

Mary, female, age 19

Part 1:

Mary is a 19-year-old woman with fragile X syndrome, moderate developmental disability, and epilepsy controlled on carbamazepine CR 400 mg BID. She takes naproxen for dysmenorrhea and lactulose on occasion for constipation. She was followed by Developmental Pediatrics and Pediatric Neurology throughout childhood, and was discharged from these services recently. She presents to your office today, accompanied by her mother, who has some concerns about her upcoming move to a group home where she will live with four other individuals with DD.

Mary is generally excited about moving out. She wants to get a job, have a boyfriend, get married, and have a baby (as her cousins have done). Her mother wants to support her wishes, but is also aware of her vulnerabilities and would like you to have a discussion with her about relationships, sexuality and parenting.

Mary's mother is also concerned about Mary's occasional difficult behaviour, and how staff may respond to this at the group home. Mary has obsessive rituals around arranging and entering her room, and becomes anxious and agitated if these are interrupted. She also becomes agitated when tired or uncomfortable, and this sometimes escalates to aggression.

How will you help Mary and her mother with the transition from pediatric care to adult care?

Part 2:

Nine months after Mary moves into the group home, the staff requests that you review her medications, because “they are not controlling her aggressive behaviour”.

Medications

- Carbamazepine CR, 600 mg BID
- Naproxen, 500 mg BID prn
- Lactulose, 15 to 30 mL BID prn
- Ovril, 21 daily x 63 days, five days off
- Lorazepam, 2 mg qam and 4 mg qhs
- Olanzapine, 2.5 mg qhs

Your records indicate you started her on Ovril at her last visit, but you notice a number of new medication changes and ask for more history.

A few months ago, at a walk-in clinic, Mary was prescribed a trial of lorazepam for insomnia. This helped for a few weeks, but then became ineffective and the dosage was increased. Despite this, her sleep did not improve, she was drowsy during the day, and her aggressive behaviour became more frequent and severe. During this time, her seizures also increased in frequency, and her neurologist increased her dose of carbamazepine CR.

Mary’s aggressive behaviour resulted in her being expelled from her work-training program and, a few weeks later, she became aggressive towards a housemate and the police were called. Mary was taken to the emergency department where olanzapine was started, and she was discharged with instructions to follow-up with her family physician.

- What, if any, changes, would you make or recommend in Mary’s medications, and how would you go about making medication changes?*
- Would knowing that Mary has fragile X syndrome influence your management of her anxiety, and choice of medications?*

Part 3:

Mary seems happy to see you, but appears tired, anxious, and is easily distracted. She has very limited eye contact and startles at sudden noises. She is shy, as usual, and keeps her coat on during the visit. She is reluctant to be physically examined and repeatedly checks her watch and says, “Time to go home.”

Physical examination

- Weight 99.5 kg; height 160 cm; BMI 39
- Heart rate 110 bpm and regular; BP 130/90 mm Hg
- Significant amount of cerumen in left ear; otitis externa in right ear
- Mid-systolic murmur
- Normal breath sounds; no adventitious sounds
- Abdomen moderately distended; non-tender; normal bowel sounds; no organomegaly
- Mild scoliosis; flat feet; laxity of joints
- Episodic staring spells; increased tone and reflexes in legs

Shortly after her visit, Mary's blood work tests positive for *Helicobacter pylori* antibodies.

- What is your differential diagnosis of what may be contributing to Mary's escalating behavioural concerns?*
- What monitoring tools would you ask group home staff to use, to help get a clearer picture of what is happening?*

Part 4:

You see Mary two weeks later. Her obsessions have become more pronounced and she explains after some prompting, "I have to sort out my room before I can sleep." Her sleep chart reveals initial insomnia and an average of four hours of sleep each night. She has difficulty sleeping because she is startled by the noises her roommate makes at night. Her sleep deprivation has resulted in a lowered seizure threshold, increased irritability and aggressive behaviour.

- What non-medication interventions would you recommend to help Mary?*
- How could the Crisis Prevention and Management Plan be used to help prevent another Emergency visit for Mary? And who should be part of the team who develops this plan?*
- If Mary has another aggressive episode and is sent to Emergency, what information would you send with her?*

Module 3 Commentary

Part 1:

❑ *How will you help Mary and her mother with the transition from pediatric care to family medicine care?*

Ideally, Mary would have started the transition process as a part of her Pediatric care at around 10 years of age, with increasing levels of information and responsibility being given to her as she entered adolescence ([CPS Position Statement on Transition to Adult Care](#)). It would be important to ask Mary and her mother what preparations they have made, and what they feel would be helpful at this point.

In general, objectives for transition include evolving self-esteem and identity, fostering independence, sexual education, achieving psychosocial stability, educational/vocational/financial planning, and healthy lifestyle promotion.

In Mary's case, this would include empowering her to speak, voice her opinion, and make choices, discussing sexual health, discussing possible arrangement of a substitute decision-maker, facilitating linkages with community services, and initiating preventive care measures (e.g., immunizations, periodic health exams, screening).

Part 2:

❑ *What, if any, changes, would you make or recommend in Mary's medications, and how would you go about making medication changes?*

Mary was prescribed lorazepam for insomnia and olanzapine for aggressive behaviour before possible causes of these behaviours were investigated. While it is not unusual to see psychotropic medications prescribed for problem behaviours in people with DD, evidence for this practice is lacking, and antipsychotic drugs such as olanzapine are not recommended for routine treatment of problem behaviours ([Consensus Guidelines #22](#)).

Isolated behaviours do not constitute a psychiatric diagnosis, and physical, environmental and emotional causes of behaviour should be investigated before psychiatric disorders are considered ([Consensus Guidelines #22](#)).

Psychotropic medication can be effective for treatment of individuals with DD who have confirmed psychiatric diagnoses, but should be used judiciously ([Consensus Guidelines #27](#)). It is important to "start low and go slow" and to monitor side effects on a regular basis. Some individuals with DD have atypical responses or side effects at low doses and may be unable to communicate this ([Consensus Guidelines #27](#)).

A trial of psychotropic medication may be considered without a psychiatric diagnosis, if careful attention is paid to the effect on target behaviours ([Consensus Guidelines #27](#)). In Mary's case, you may wish to consider reducing or stopping olanzapine on a trial basis to see if there is any change in behaviour ([Consensus Guidelines #22](#)). If no change is appreciated, it could be discontinued.

Antipsychotic medications can have serious side effects and can increase risk of metabolic syndrome ([Consensus Guidelines #28](#)), so there is good reason to discontinue them if they have not having the desired effect.

The [Auditing Psychotropic Medical Therapy](#) tool is available to aid in this decision. Further guidelines on prescribing psychotropic medications can be found in the [Psychotropic Medication Issues](#) tool.

❑ *How would knowing that Mary has fragile X syndrome change your management of her anxiety, and choice of medications?*

Some syndromes, such as fragile X, are associated with behavioural phenotypes in which certain developmental, neurologic or behavioural manifestations are more likely to be present ([Consensus Guidelines #23](#)), and this can influence medication choice. Aspects of the fragile X syndrome behavioural phenotype include aggressive behaviour, sensory defensiveness, ADHD, mood instability and anxiety ([Fragile X Syndrome Health Watch Table](#)).

For fragile X patients with ADHD, non-stimulant medication is often a better choice than stimulant medication, which can cause increased anxiety, irritability, and labile mood. Benzodiazepines should be used cautiously in people with fragile X syndrome because they sometimes have paradoxical reactions to this class of medication, resulting in behavioural dis-inhibition, irritability, and aggression. SSRIs have been widely used for anxiety, with mixed results. There is some evidence for Aripiprazole in the treatment of irritability [Tranfraglia 2011].

A diagnosis of autism should also be considered for Mary, as 30% of individuals with fragile X syndrome have this comorbidity ([Fragile X Syndrome Health Watch Table](#)). Management of what appear to be social anxiety and OCD traits may change in light of this diagnosis.

Part 3:

❑ *What is your differential diagnosis of what may be contributing to Mary's escalating behavioural concerns?*

There are a number of different issues that may be contributing to the changes in Mary's behaviour. [A Guide to Understanding Behavioural Problems and Emotional Concerns](#) can be used to aid with diagnostic formulation.

On review of possible medical conditions that could be contributing to the behavioural exacerbation, likely possibilities would include constipation, dysmenorrhea, adverse reaction to new oral contraceptive pill, and seizures. Based on her exam, we also know she has otitis externa, which would be painful and cerumen blockage, which may be contributing to hearing loss. She has also tested positive for H. pylori, which can be associated with peptic ulcer disease.

The [Fragile X Syndrome Health Watch Table](#) highlights a number of other possible medical conditions commonly associated with the syndrome, including obstructive sleep apnea, recurrent UTIs, joint dislocations, and premenstrual symptoms.

Environment supports and expectations should also be taken into consideration. The changes in Mary's living environment and routine may have been stressors contributing to her behaviour change. It would also be important to determine whether staff at the group home is over- or under- estimating her abilities, whether her needs are being met, and whether caregivers feel adequately supported and trained to care for her.

Emotional issues may be contributing to Mary's behavioural changes as well. She has just gone through a major life transition (moving away from the family home) and is navigating new relationships. She may also be experiencing new insight into how her disability could impact her life. Caregivers and family members may be able to assist in determining whether emotional factors are at play.

Psychiatric disorders are more common among adults with DD as compared to the general population, and care should be taken to recognize them. Tools such as the Aberrant Behaviour Checklist-Community (ABC-C) and Psychiatric Assessment Schedule for Adults with DD (PAS-ADD) are available to aid in recognizing psychiatric disorders in the DD population ([Consensus Guidelines #23](#)). ADHD and anxiety disorders are common in fragile X syndrome and Mary has displayed some OCD tendencies. Special attention should be given to these possibilities.

❑ *What monitoring tools would you ask group home staff to use to help get a clearer picture of what is happening?*

In order to understand the basis for problem behaviours and to treat them effectively, it is important to seek input from adults with DD and their caregivers ([Consensus Guidelines #25](#)). Target symptoms (e.g., sleep, seizure activity, bowel habits, menses) and behaviours (e.g., physical and verbal aggression, self-injury, damage to environment) should be identified and monitored using available tools.

Monitoring tools are available on the Surrey Place Website in the [Tools for Caregivers](#) section.

A chart to monitor daily functions is also available in the [Today's Visit](#) Tool and an [Antecedent-Behaviour-Consequence Chart](#) is available for analysis of possible behavioural triggers.

Part 4:

❑ *What non-medication interventions would you recommend to help Mary?*

Non-pharmacological interventions are often effective in the prevention and treatment of problem behaviours ([Consensus Guidelines #26](#)). In general, interventions could include addressing sensory issues, environmental modification, education and skill development, communication aids, psychological and behavioural therapies, and caregiver support.

In Mary's case, environmental modification to improve her ability to sleep soundly could potentially have a positive effect on her behaviour. Ideas include moving Mary to a different room, earplugs, a white-noise machine, or addressing her roommate's sleeping issues.

Other interventions for Mary could include CBT to address anxiety and OCD tendencies, and social skills training around effective communication and personal boundaries awareness.

Education for her caregivers on fragile X syndrome would also be appropriate.

❑ *How could the Crisis Prevention and Management Plan be used to help prevent another Emergency visit for Mary? And who should be part of the team who develops this plan?*

Sometimes behavioural crises, such as the one Mary experienced, require management in an emergency department. If an adult with DD is at high risk for recurrent crises, a crisis plan should be developed ([Consensus Guidelines #29](#)). A template and instructions for developing a *Crisis Prevention and Management Plan* can be found on pages 77-81 of the Tools for Primary Care of People with DD.

This type of plan outlines recommended caregiver responses corresponding to different stages of escalating patient behaviour. Positive approaches and structured routines are used to encourage calm behaviour. Supportive talk and environmental modification (e.g., decreased noise and increased personal space) are used to prevent anxiety. Provision of direction (e.g., generating discussion, demonstrating understanding, providing reassurance, setting limits) and further environmental modification are potential responses to escalating behaviour.

Safety strategies (e.g., removing potentially harmful objects, maintaining space, reminding about boundaries) and crisis response strategies (as agreed upon by the care team) are used to respond to crisis behaviours. A post-crisis resolution strategy is also an important part of the plan.

When possible, a case manager or behavioural therapist would be an ideal person to lead the development of a [Crisis Prevention and Management Plan](#). A team approach to planning should be taken and should involve the person with DD, their caregivers,

health care professionals involved in their care, a service coordinator and, in certain circumstances, Emergency Services (e.g., police, paramedics, Emergency Department staff).

Once established, the plan should be reviewed on a regular basis.

❑ *If Mary has another aggressive episode and is sent to Emergency, what information would you send with her?*

If Mary were to be sent into the Emergency Department again, an [Essential Information for Emergency Department \(ED\)](#) form could be completed. This form includes patient identification and contact information for caregivers, Substitute Decision-Maker, and health care professionals involved in care. It identifies the reason for referral to the ED and identified safety risks. (The [Risk Assessment Tool for Adults with DD in Behavioural Crisis](#) can be used to help identify risks).

It also provides a brief overview of Health Status, which in Mary's case would include her diagnosis of moderate DD secondary to fragile X syndrome, epilepsy, and history of aggression with underlying anxiety. Special needs are also identified, which in Mary's case include sensitivity to noise, difficulty with eye contact, and hesitancy around physical examination.

In addition to the *Essential Information for ED* form, a list of current medications, information on PRN medication given, and a copy of the *Crisis Management and Prevention Plan* should also be sent to the Emergency Department.

References for Module 3 of Part 3: Case Modules

1. Canadian Pediatric Society. (2007) "Transition to adult care for youth with special health care needs". Position Statement (AH 2007-01). *Pediatric Child Health*, 12: 785-88.
2. Sullivan et al. [Primary Care of adults with developmental disabilities. Canadian Consensus Guidelines](#). *Can Fam Physician* 2011;57: 541-53.
3. Tranfaglia et al. (2011) "The Psychiatric Presentation of Fragile X: Evolution of the Diagnosis and Treatment of the Psychiatric Comorbidities of Fragile X Syndrome". *Dev Neurosci* 33: 337-48.
4. Developmental Disabilities Primary Care Initiative. *Tools for the Primary Care of People with Developmental Disabilities*. 1st ed. Toronto: MUMS Guideline Clearinghouse; 2011.

Discussion Starters

This module contains short clinical scenarios that aim to draw the learner's attention to a specific Guideline or Tool from the Primary care of adults with developmental disabilities: Canadian consensus guidelines.

Their intended use is either for a facilitator to present to learners to stimulate a discussion, or for an individual learner to go through a shorter exercise in familiarizing themselves with the Tools and Guidelines.

After reading each case, the learner should reflect on any further questions they may have in the patient's history, any investigations they might like to order, and any preventive care strategies from which the patient would benefit.

Case A: Respiratory Disorders

Karen is a 65-year-old woman with cerebral palsy, spastic quadriplegia, seizure disorder and a severe intellectual disability. She lives in a group home. Over the past year, she has had four episodes of pneumonia which required hospitalization and which is hypothesized to be due to aspiration.

Staff at her group home have also noted that it is taking much longer to feed her (between 60 to 90 minutes) and that at times she refuses oral intake altogether. She has been slowly losing weight and now has a BMI of 16.

1. What further information would you like from the patient's history?
2. Are there any investigations you would like to order?
3. Are there any preventive care strategies from which the patient would benefit?

Discussion A:

Karen should be referred for a video fluoroscopy swallowing study to assess her safety to take foods by mouth, and considered for a G-tube if the results show that she is aspirating. Respiratory disorders like aspiration pneumonia are among the most common cause of death for adults with developmental disabilities.

Swallowing difficulties are prevalent in those patients with neuromuscular dysfunction or taking certain medications with anticholinergic side effects, and they may result in aspiration or asphyxiation. Patients with developmental disabilities should be screened for possible signs of swallowing difficulties at least annually to rule out dysphagia and aspiration, noting increased throat swallowing after coughing, choking, drooling, long meal times, aversion to feeding, weight loss and frequent chest infections ([Consensus Guidelines #14](#)).

The Cerebral Palsy Health Watch Table (available soon on the Surrey Place website) could also be useful to Karen's family physician for ongoing monitoring of her health.

Case B: Cancer Screening

Monica is a 60-year-old woman with cerebral palsy, seizure disorder, gastroesophageal reflux disorder and a mild to moderate developmental disability. She has lived at home with her sister since her mother died ten years ago. She attends a supported employment program and goes swimming once a week.

She has monitoring blood work done every six months. You notice that her hemoglobin has slowly been dropping. One year ago it was 135, six months ago it was 120, and now it is 105. You confirm that she has an iron deficiency with a ferritin of 5.

A colonoscopy is ordered, which finds a large mass at the recto-sigmoid junction. The mass is surgically removed that week and she is referred to medical and radiation oncologists for further treatment.

1. What further information would you like from the patient's history?
2. Are there any investigations you would like to order?
3. Are there any preventive care strategies from which the patient would benefit?

Discussion B:

Cancer screening is an important aspect of preventive care. Unfortunately, adults with developmental disabilities are less likely than those in the general population to be included in preventive screening programs, such as cervical screening, breast examination, mammography and digital rectal examination.

In Karen's case, given that she is 60 years old, she should already have had screening for breast cancer, colon cancer and cervical cancer if she has a history of sexual activity.

Patients with developmental disabilities are also less likely or able to do self-examination or to report abnormalities, such as noticing a breast mass or a change in stool caliber. It is therefore important for someone else in their lives to monitor for these symptoms, for family physicians to be more intensive with physical examination, and for screening to be done regularly.

This can, however, be challenging; for example, performing mammography in a woman with skeletal malformations who cannot sit upright. In this case, it may be worth exploring alternatives such as clinical breast exam or ultrasound.

Of note, colorectal cancer risk is greater for women than for men with developmental disabilities. Patients with developmental disabilities should receive regular cancer

screening for cervical, breast, testicular, prostate and colon cancer following the same guidelines as the general population ([Consensus Guidelines #21](#)).

Case C: Musculoskeletal Disorders

Simon is a 37-year-old man with an intellectual disability in the severe range and a seizure disorder. He lives in a group home with five other residents. He ambulates but is at times unsteady and has a history of falls. He has no known allergies, and has been on carbamazepine and valproic acid since he was a child for his seizures. He has a seizure once every three to four months.

Yesterday, Simon had a seizure which caused him to fall on his outstretched right wrist. After recovering from the seizure, his group home staff found that his wrist was very swollen and that he wasn't using it. An x-ray at the hospital showed a Colles' fracture of his right wrist.

1. What further information would you like from the patient's history?
2. Are there any investigations you would like to order?
3. Are there any preventive care strategies from which the patient would benefit?

Discussion C:

Osteoporosis and osteoporotic fractures are more prevalent and tend to occur earlier in adults with DD than in the general population. Simon should be screened beginning in early adulthood, given his risk factor of long-term anticonvulsant use.

There are several risk factors for osteoporosis and fragility fractures that are common in patients with developmental disabilities. In addition to aging and menopause in women, consider the severity of the developmental disability, low body weight, reduced mobility, increased risk of falls, smoking, hypogonadism, hyperprolactinemia, the presence of particular genetic syndromes (e.g., Down syndrome, Prader-Willi), and long-term use of certain drugs (e.g., glucocorticoids, anticonvulsants, injectable long-acting progesterone in women) as important risk factors for osteoporosis.

In addition to early screening, Simon should be started on calcium and vitamin D supplementation to prevent further bone loss. Now that he has had a fragility fracture, he should be considered for initiation of bisphosphonate therapy as long as there is no co-morbid gastroesophageal reflux disease. Initiation of bisphosphonates in young patients with developmental disabilities is controversial, as there is little research outlining its positive and negative effects in long-term use ([Consensus Guidelines #17](#)).

Case D: Psychotic Disorders

James is a 32-year-old man with Down syndrome, an intellectual disability in the moderate to severe range, and hearing impairment. He currently lives at home with his mother and attends a day program during the week. James has limited expressive language, but has always engaged in self-talk, which was determined through psychological assessment at age 15 to be adaptive and self-soothing in nature.

Recently, however, the frequency of self-talk has increased significantly. Furthermore, it has changed in character from quiet and soothing to loud with yelling outbursts. His words are not clear enough to determine the content of his self-talk.

James's mother brings him to see you today because she is worried that he is hallucinating and is requesting that he be started on a medication.

1. What further information would you like from the patient's history?
2. Are there any investigations you would like to order?
3. Are there any preventive care strategies from which the patient would benefit?

Discussion D:

Given the change in character of James's self-talk, his mother is right to be concerned. As with any behaviour change, underlying medical conditions, problems with supports and expectations, and emotional issues must be ruled out before diagnosing a psychiatric disorder.

Despite James's mother's request for a medication, unless there is an acute behavioural crisis, medication is not the first line until a psychiatric diagnosis is made. Psychotic disorders are very difficult to diagnose when delusions and hallucinations cannot be expressed verbally, as in James's case. He may be experiencing developmentally appropriate self-conversation and not a hallucination.

If you are concerned that your patient with a developmental disability may have developed a psychotic disorder, seek interdisciplinary input from specialists in psychiatry, psychology and speech-language pathology with expertise in developmental disabilities, or in this case, Down syndrome, to help clarify diagnoses in patients with limited or unusual use of language.

As depression is common in Down syndrome, especially around times of transition, a mood disorder should also be ruled out ([Consensus Guidelines #24](#)).

Case E: Prader-Willi Syndrome Health Watch Table

Julian is an 18-year-old man with Prader-Willi syndrome (PWS). He presents to your office as a new patient with his parents. He lives at home with his mother, father and his 16-year-old sister. He has not seen a physician since he was twelve years old.

He is here today with a cough and dyspnea. Julian is obese with a BMI of 32. His family is struggling to help him adhere to his strict calorie-restricted diet, as he frequently finds ways to steal food.

1. What further information would you like from the patient's history?
2. Are there any investigations you would like to order?
3. Are there any preventive care strategies from which the patient would benefit?

Discussion E:

The Prader-Willi Syndrome Health Watch Table can help you to focus on your priorities for your new patient, Julian. PWS has identified health concerns that all people with PWS should be screened for. For example, people with PWS tolerate upper respiratory infections poorly, and he should therefore be assessed and treated as soon as possible for his symptoms of URI today.

Since Julian hasn't seen a doctor in many years, it would be a good opportunity to discuss other preventive care issues for people with PWS, including dental, vision and hearing screening, cardiac evaluation, sleep study, and bone mineral density.

Blood work should be done to screen for diabetes, hyperlipidemia, thyroid dysfunction ([Prader-Willi Syndrome Health Watch Table](#)).

Julian's family should also be supported through this difficult time and connected with the available local resources that may help them. You may want to see Julian and his family at an increased frequency, advocate for respite care if the funding is available, or refer his parents for counselling, if they are interested.

References for Discussion Starters of Part 3: Case Modules

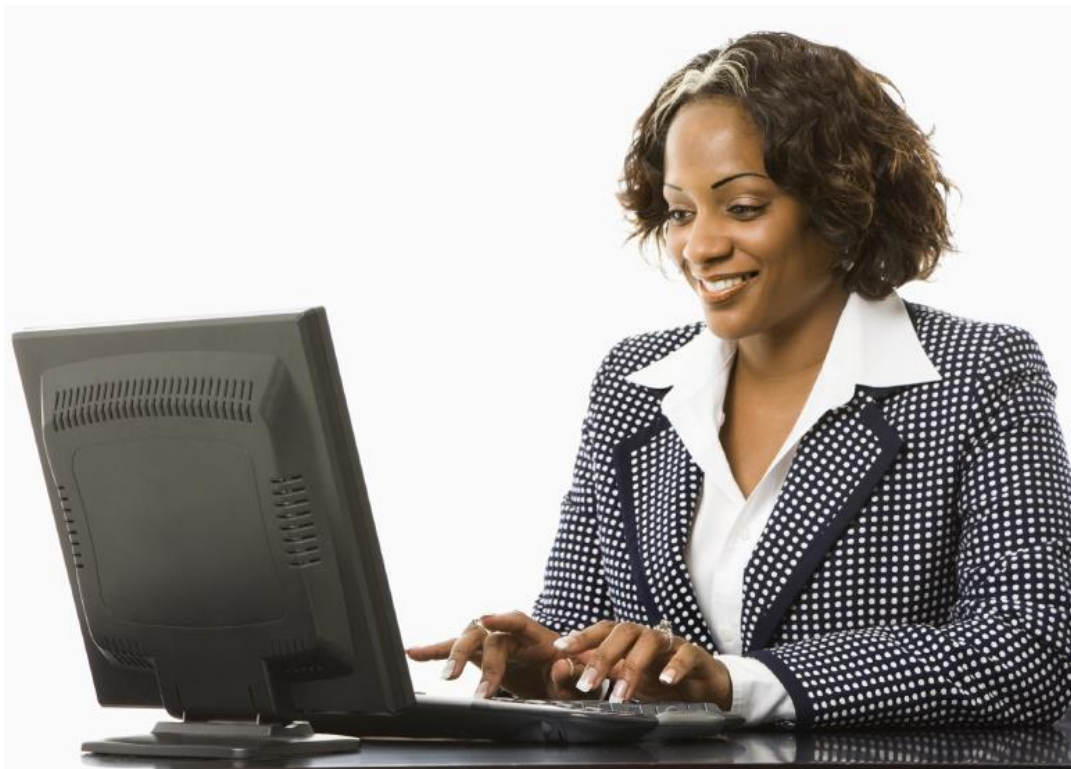
1. Sullivan et al. [Primary Care of adults with developmental disabilities. Canadian Consensus Guidelines](#). Can Fam Physician 2011;57: 541-53.
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***Suggested citation for Part 3: Case Modules:**

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Family Medicine Curriculum Resource: Adults with Developmental Disabilities

Part 4: Resident Assessment



Family Medicine Curriculum Resource: Adults with Developmental Disabilities

Part 4 – Implementing DD Curriculum and Assessing Resident Competencies in a Teaching Practice*

1. Implementation of Curriculum through Clinical Encounters

Clinical encounters with an adult with a developmental disability are the most important way to learn and to assess residency level competencies in DD, especially if the resident is prepared for the encounter; for instance, through reading:

- [Primary care of adults with developmental disabilities: Canadian consensus guidelines](#).
- The Primer (Part 2 of this Curriculum Resource) which frames the [Guidelines and Tools](#) in the context of the competencies for Family Medicine Residents (Part 1 of this Curriculum Resource).
- The case modules (Part 3 of this Curriculum Resource), which can be used for self-learning or small group learning. The case modules are based on the Guidelines and illustrate use of the Tools.

Residents' learning through clinical encounters could be opportunistic – when patients seek care for specific symptoms – or it could be through performing a comprehensive health assessment or periodic health review (i.e., a periodic health exam, annual physical or health check). A comprehensive health assessment is a way to integrate education and resident assessment with best clinical practices, because good evidence [1-3] supports comprehensive health assessments for persons with DD as a way to increase health promotion, preventive maneuvers and case-finding.

- The [Cumulative Patient Profile](#) and/or the Preventive Care Checklist for [males](#) or [females](#) adapted for persons with DD can be used to guide and record a comprehensive health assessment.
- The Guidelines and other Tools can be used to provide “point-of-care” education through annotations on a medical record form, similar to the explanations available for items on the “Rourke Record” (an example of such a DD form is available from the Department of Family Medicine, Queen’s University).

Steps for planning a comprehensive health assessment could include the following:

1. Identify an adult with DD in the practice and invite for a checkup, if appropriate, with caregiver or substitute decision-maker. Identify the best time of day or other accommodations needed to prepare for the visit.
2. Request the patient/caregiver fill out the [Caregiver Health Assessment](#) form or [Today's Visit](#) form in advance, to bring to the appointment for background information.
3. Seek past medical or school records to obtain the most recent functional and genetic assessments.
4. Involve other members of the inter-professional health team to collect information to complete parts of the [Cumulative Patient Profile](#) and/or the Preventive Care Checklist for [males](#) or [females](#).
5. Complete the health assessment with a physical exam, development of a problem list and a plan for treatment and follow up of issues identified. Expect that more than one visit will be necessary to accomplish the whole process.

2. Assessment of Resident Competencies by Observation of Clinical Activities

The observation of clinical activities in practice allows a teacher (or resident) to infer the presence of competencies [4].

The rubric in the Appendix to this section can identify the progress of residents to the competencies listed in Part 1 of this Curriculum Resource.

- In the columns of the rubric, increasing levels of performance are identified by the degree of supervision of the resident necessary by the teacher. Naming the levels in this way reflects the practical, somewhat intuitive assessments made by clinical teachers in the course of patient care in their teaching practices:
 - Close Supervision
 - Minimal Supervision
 - Supervision for Refinement only (i.e., competence at the level of a resident ready to enter practice)
- In the rows, the rubric is organized by phases of the clinical encounter and by one of the DD competencies, so a clinical teacher observing a resident for a few minutes in one part of the clinical encounter can easily identify a competency to assess in that phase of the encounter.

References for Part 4: Resident Assessment

1. Lennox N, Bain C, Rey-Conde T, Purdie D, Bush R, Pandeya N. Effects of a comprehensive health assessment programme for Australian adults with intellectual disability: a cluster randomized trial. *Int J Epidemiol* 2007;36(1):139-46.
2. Lennox N, Ware R, Bain C, Taylor Gomez M, Cooper S. Effects of health screening for adults with intellectual disability: a pooled analysis. *Br J Gen Pract* 2011;61:193–6.
3. Robertson J, Roberts H, Emerson E, Turner S, Greig R. The impact of health checks for people with intellectual disabilities: a systematic review of evidence. *Journal of Intellectual Disability Research* 2011; 55(part II): 1009–19.
4. ten Cate O, Scheele F. Competency-based postgraduate training: can we bridge the gap between theory and clinical practice? *Academic Medicine* 2007;82(6):542-7.

***Suggested citation:**

Casson I, Grier E, Griffiths J. Resident Assessment. In: Curriculum Working Group, Developmental Disabilities Primary Care Initiative. [Family Medicine Curriculum Resource: Adults with Developmental Disabilities](#) [Internet]. Toronto (ON): Surrey Place Centre; c 2009 [updated 2014 June; cited 2014 Feb 8]. [pp. 75-81].

Rubric for Assessing DD Competencies in Clinical Encounters

Note: Numbers assigned to competencies correspond to those in Part 1 of the Family Medicine Curriculum Resource.

Phase of the clinical encounter observed	DD Competency that can be assessed	Level of performance (expressed as level of supervision required)		
		Close Supervision	Minimal Supervision	Supervision for Refinement Only (Competent)
History	<p>Demonstrates respect and empathy (Competency 1a)</p> <p>Communicates effectively (2a)</p>	<ul style="list-style-type: none"> • Unable to adjust communication methods to accommodate patients with intellectual or communication disability. • Speaks with caregiver instead of patient. • Does not confirm understanding. • Speaks too loudly, does not allow for assimilation of information. 	<ul style="list-style-type: none"> • Attempts to adjust communication methods to accommodate patients with intellectual or communication disability. • Attempts the appropriate pacing of information, but not able to be completely successful such that assimilation of information and understanding is ensured. 	<p>1a) Demonstrates respect and empathy for patients with developmental disabilities as individuals (e.g., in the way the resident includes the person with DD in the clinical encounter, rather than speaking about him/her to the caregiver.</p> <p>2a) Communicates effectively, applying communication methods in accord with the patient's individual expressive and receptive communication capabilities (e.g., simple language, writing and pictures, taking extra time).</p>

Phase of the clinical encounter observed	DD Competency that can be assessed	Level of performance (expressed as level of supervision required)		
		Close Supervision	Minimal Supervision	Supervision for Refinement Only (Competent)
Physical examination	Adjusts the clinical environment (2b)	<ul style="list-style-type: none"> Makes no special accommodation around the physical exam. 	<ul style="list-style-type: none"> Attempts to make accommodation around the physical exam to make the process easier, safer and more accessible. 	<p>2b) Adjusts the clinical environment (e.g., removes barriers to physical exams) for patient comfort, taking into account sensory defensiveness (e.g., annoyed by bright lights and loud noises). Considers home visit to observe patient in their own environment and to do the physical exam, if appropriate.</p>
Investigation	Etiology and functional ability (3)	<ul style="list-style-type: none"> Unaware that etiologic or functional assessment are important in the care of this patient. 	<ul style="list-style-type: none"> Begins to think through some helpful investigations, but knowledge is insufficient to arrange or to be comprehensive or to fully manage the investigations. 	<p>3a) Uses etiologic information (e.g., a diagnosed genetic syndrome, like Down syndrome) for anticipatory and preventive care and for acute presentations. When etiologic information is not available, assesses the need for further investigation (e.g., referral for genetics assessment or neuroimaging). 3b) Uses (or obtains, if necessary) up-to-date assessments of intellectual and functional abilities (e.g., psychological, educational or</p>

Phase of the clinical encounter observed	DD Competency that can be assessed	Level of performance (expressed as level of supervision required)		
		Close Supervision	Minimal Supervision	Supervision for Refinement Only (Competent)
				vocational assessments) to determine appropriate expectations in clinical encounters and the adequacy of supports.
Diagnosis	Recognizes illnesses commonly associated with DD and the atypical presentations of serious illness (4)	<ul style="list-style-type: none"> • Not aware of syndrome-specific issues. • Does not consider full range of possible contributors when dealing with changes in behaviour. • Does not have a holistic approach. • Is not able to demonstrate knowledge of how a Developmental Disability may impact the possible differential diagnoses. 	<ul style="list-style-type: none"> • Aware of some syndrome-specific issues that are clinically relevant. • Considers some, but not the full range of possible contributors, when dealing with changes in behaviour. • Has a holistic approach, but does not fully appreciate the impact of the Developmental Disability on the possible differential diagnoses. 	<p>4a) In generating differential diagnoses and management plans, demonstrates knowledge of the frequency, preventability, and treatability of various health issues specific to adults with DD (e.g., the most common cause of death is respiratory disease; common illnesses associated with DD are autism, epilepsy, cerebral palsy, mental health problems).</p> <p>4b) Recognizes atypical presentations of serious illness (e.g., behaviour change as a presentation of reflux esophagitis) and manages appropriately.</p>

Phase of the clinical encounter observed	DD Competency that can be assessed	Level of performance (expressed as level of supervision required)		
		Close Supervision	Minimal Supervision	Supervision for Refinement Only (Competent)
Treatment and Management	Ethics – Facilitates consent (1b) and supports substitute decision-makers (1c)	<ul style="list-style-type: none"> Does not understand the issues of consent and capacity. Does not establish who the substitute decision-maker is. Makes no special accommodation necessary for a procedure. 	<ul style="list-style-type: none"> Attempts to deal with capacity issues and obtain consent, but either misjudges capacity or is unable to obtain consent. Attempts to make accommodation for a procedure to make the process easier, safer and more accessible. 	<p>1b) Facilitates informed consent in persons with partial decision-making capacity (e.g., by asking if they would normally ask for help in making similar decisions).</p> <p>1c) Identifies and supports substitute decision-makers (e.g., in using best practices as a guide to decision making).</p>
Follow up	Facilitates access to other sources of care (2c)	<ul style="list-style-type: none"> Little attempt for continuity of care. Does not incorporate others in the follow up. 	<ul style="list-style-type: none"> Attempts continuity of care, and may understand the significance of this, but may not plan adequately for anticipatory care. Some useful referrals made. 	<p>2c) Facilitates access to the family practice team, consultants, hospital services (e.g., planning visits to imaging departments) and community resources (e.g., completing disability applications, directing to developmental services in the social and educational sectors).</p>