

❖ Health Care Maintenance Guidelines for Individuals with Developmental Disabilities

Introduction

The following tables were developed to assist primary care providers, patients, families and caregivers to understand the complexities of health screening. Individuals with developmental disabilities (DD) are living longer and in many instances have a life expectancy similar to those in the general population. Significant disparities exist for people with developmental disabilities. People with developmental disabilities have difficulty accessing medical care for chronic health conditions as well as preventative health screening and counseling.¹

While there are no large scale population-based studies of screening and prevention for individuals with DD, there have been a number of consensus statements and guidelines recently developed to help guide primary care providers' decision-making.

The recommendations in the following tables were compiled and adapted from a review of the literature that includes major national professional organizations and societies (ACS, AAFP, ACOG, USPSTF, etc.) as well as specialty DD organizations such as AAIDD, consensus statements and expert clinical opinion. Many of the recommendations are not evidenced-based but represent a reasonable approach to testing. Some of the guidelines here recommend testing at "regular intervals." Unfortunately there is no consensus about the frequency of screening intervals for certain tests so the term "regular intervals" is used to capture these uncertainties.

Compared to the general population, individuals with developmental disabilities *require a more proactive approach* to health care maintenance. The highlighted areas in the tables are health problems that are seen more frequently in this population and require additional attention by health care providers.

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¹ Horwitz SM, Kerker BD, Owens PL and Zigler E. The health status and needs of individuals with mental retardation. Department of Epidemiology and Public Health, Yale University School of Medicine; (2000) Department of Psychology, Yale University, New Haven, CT. Available at: <http://info.specialolympics.org/Special+Olympics+Public+Website/English/Initiatives/Research/Health+Research/Health+Status+and+Needs.htm>. Accessed March 8, 2010.

HEALTH CARE MAINTENANCE GUIDELINES FOR ADULTS WITH DEVELOPMENTAL DISABILITIES

NOTE: Health problems that are seen more frequently in this population and require additional attention by health care providers are highlighted in the darker shade of green.

HEALTH PROBLEM	19 – 40 YEARS	40 – 65 YEARS	65 YEARS AND OLDER
►Abuse & Neglect	• Identify and evaluate unexplained physical and/or behavioral signs and symptoms at each visit. These signs might include some of the following: unexplained bruising, falls, injuries, weight loss, depression and behavior changes.		
Alcohol & Substance Abuse	• Screen yearly.		
Breast Cancer (Women)			
Clinical Breast Exam	<ul style="list-style-type: none">• Perform yearly breast exam every two to three years.⁴• Routine screening with clinical breast exam not recommended.²²	<ul style="list-style-type: none">• For ages 40-65 screen with clinical breast exam yearly.⁴• Start screening at age 50 with yearly clinical breast exam.²²	<ul style="list-style-type: none">• Perform clinical breast exam every one to two years if life expectancy is greater than or equal to five years.²¹
Mammography	<ul style="list-style-type: none">• Routine screening with mammography not recommended.²²	<ul style="list-style-type: none">• Order screening mammography yearly.⁴• Women ages 40-49 routine mammogram not recommended.²²	<ul style="list-style-type: none">• Order screening mammography every two years until age 74. At age 75, consider stopping routine screening if patient has significant medical problems that threaten life expectancy.²²• Order screening mammography every one to two years if life expectancy is greater than or equal to five years.²¹
BRCA	<ul style="list-style-type: none">• For women with a family history associated with increased risk for BRCA1 or BRCA2 gene mutations, consider referral for genetic counseling and evaluation.²⁰		--
<div>Comments</div> <ul style="list-style-type: none">-Family history is often difficult to obtain in this population. Individual decision-making is critical. Inform women and caregivers of potential benefits and consequences of breast cancer screening.-Consider clinical breast exam as women with intellectual disabilities may not understand the significance of changes or have the skills to communicate changes they notice. Also, women with sensory or neuromuscular problems may have difficulty performing an accurate self-exam.-Consider genetic testing for the BRCA-A gene for women who have a strong family history and are unable to do a mammogram.²⁰			
►Cervical Spine Atlanto-Axial Instability	<ul style="list-style-type: none">• Perform an annual neurologic examination for signs and symptoms of spinal cord injury for patients with Down syndrome.¹• Order Cspine x-ray with lateral flexion and extension if symptoms develop, such as: changes in behavior or activity, changes in hand preference or urinary incontinence. If this is the first Cspine, also order an anteroposterior view.		
<div>Comments</div> <ul style="list-style-type: none">-Consider screening cervical spine films prior to participation in athletics.			
Cervical Cancer (Women)	Individualized decision-making depending on patient risk and sexual history. No consensus, but major groups recommend the following:		
	<ul style="list-style-type: none">• Before age 21, avoid pap smear screening regardless of sexual activity. At age 21, begin pap smear screening. Ages 21-29 screen every two years, then every three years for women with three consecutive normal paps.³• Perform pap smear within three years after first sexual intercourse or by age 21, whichever comes first.^{22, 19}	<ul style="list-style-type: none">• Perform pap smear screening every three years for women with three consecutive normal paps.³• Perform pap smear screening every one to three years.^{22, 19}	<ul style="list-style-type: none">• At age 65²², or age 70⁴, consider discontinuing pap smear screening if the patient has had three or more documented, consecutive normal tests.
<div>Comments</div> <ul style="list-style-type: none">-See Breast Health Access for Women with Disabilities (BHAWD): “Table Manners and Beyond: The Gynecological Exam for Women with Developmental Disabilities and Other Functional Limitations”: www.bhawd.org/sitefiles/TblMrs/cover.html-See “Tips for a Successful Pelvic Exam”:- http://familymedicine.medschool.ucsf.edu/odpc/docs/pdf/practice_pearls/tips_for_a_successful_pelvic_exam.pdf			

HEALTH PROBLEM	19 – 40 YEARS	40 – 65 YEARS	65 YEARS AND OLDER
Chlamydia	<ul style="list-style-type: none"> Screen annually for sexually active women through age 26.⁶ 	<ul style="list-style-type: none"> For patients age 40 and older, since there is no data, individualized decision-making is appropriate. 	
	Comments <i>-Patients may not reliably report sexual activity or symptoms.</i>		
Cholesterol & Lipid Disorders	<ul style="list-style-type: none"> Order a fasting lipid panel every five years. More frequently if patient is taking atypical antipsychotic medications or has diabetes.²² 		
Colorectal Cancer	--	<ul style="list-style-type: none"> At age 50, screen with one of the following strategies: <ol style="list-style-type: none"> Colonoscopy every ten years. Flexible sigmoidoscopy every five years. Fecal Occult Blood Test (FOBT) annually plus flexible sigmoidoscopy every three years. FOBT annually. (UPSTF 2002, ASGE 2006, AAFP 2008).¹⁰ 	
	Comments <i>-Depending on the patient's co-morbidities, anesthesia risk may outweigh the benefits of colonoscopy.</i> <i>-Patients with mobility disorders, spasticity, and/or cognitive impairment may require hospital admission the day prior to testing with colonoscopy and sigmoidoscopy for professional assistance with the bowel preparation.</i>		
►Dental Disease	<ul style="list-style-type: none"> Perform an annual oral exam. Refer to dentist for regular dental care including cleaning every six months or as recommended by the dentist. Pay special attention to dental and gum health in persons with certain syndromes, such as Cornelia de Lange, Cerebral Palsy, Down, Prader-Willi, Turner, Rett, Williams and Tuberous Sclerosis.¹⁶ 		
	Comments <i>-Patients with developmental disabilities are at high risk for periodontal disease for numerous reasons, including: difficulty maintaining hygiene, lack of access to regular dental care, syndrome-specific susceptibilities and medications.</i> <i>-In some patients unable to tolerate office exams and treatment, hospital dentistry under anesthesia may be indicated. Other necessary diagnostic testing should be considered while patient is sedated.</i>		
►Depression	<ul style="list-style-type: none"> Screen annually or sooner for behaviors or emotions that may indicate depression.¹⁶ 		
	Comments <i>-Patients with developmental disabilities may have difficulty recognizing and communicating symptoms such as depressed mood, anxiety, and sadness. Mental health symptoms are often expressed in physical or behavioral changes. It is critical that health care providers obtain information about the patient's usual level of functioning, skills and behavior in order to assess the potential for mental health disorders.</i> <i>-See Diagnostic Manual-Intellectual Disabilities for more in-depth discussion on assessment.</i> ¹¹		
►Diabetes	<ul style="list-style-type: none"> Screen at least every three years until age 45. Screen annually after age 45 and annually for patients on antipsychotic medication and those with syndromes associated with diabetes, such as Prader-Willi, Klinefelter, Turner and Down.¹⁶ 		
	Comments <i>-Individualized decision-making about screening is appropriate for other individuals with developmental disabilities.</i> <i>-See American Diabetes Association. Consensus Statement on Antipsychotic Drugs and Obesity and Diabetes:</i> http://care.diabetesjournals.org/content/27/2/596.full?ijkey=8499e4a1017d5491d7d7bb66d30b77013a2397c8&keytype2=tf_ipsecsha		
Fall Risk	<ul style="list-style-type: none"> For all ages: evaluate as part of the annual physical examination including an evaluation of the medication profile for drugs that may impact balance and/or gait. Screen more frequently if there is a change in gait/balance or for individuals at high risk, such as those who have a history of two or more falls in the previous year.¹⁶ For patients with no previous mobility impairments who report one or more falls, consider performing the "Get-Up and Go Test": http://www.ncbi.nlm.nih.gov/pubmed/3487300. Patients having difficulty with this test should be referred to a physical/occupational therapist for a full fall evaluation. If the patient has had an increase in falls or a decline in function, a medical evaluation of the cause is warranted. 		
HIV	<ul style="list-style-type: none"> For ages 13-64 screen with at least one HIV test in their lifetime. Test periodically for patients at-risk (sexually active without barrier protection, multiple partners, men who have sex with men, all pregnant women, history of sexually transmitted diseases).^{6, 22} 		--

HEALTH PROBLEM	19 – 40 YEARS	40 – 65 YEARS	65 YEARS AND OLDER													
►Hearing	• Screen annually subjectively or objectively with office-based testing (Whisper Test).															
	--	• Refer to audiology at regular intervals. • Refer to patients for hearing assessment every five years after age 50 (every three years throughout life for patients with Down syndrome). • Reevaluate hearing if problems are reported or changes in behavior are noted. ¹⁷														
	Comments -Other syndromes associated with hearing impairments include Cornelia de Lange, Noonan, Usher, and Smith-Magenis. ¹⁶ -Methods for testing may include the following:															
	<table><tr><th>Method</th><th>Applicable for Developmental Age (years)</th></tr><tr><td>OtoAcoustic Emissions (OAE)</td><td>> 0</td></tr><tr><td>Auditory Brainstem Responses (ABR)</td><td>> 0</td></tr><tr><td>Behavioural observation audiometry</td><td>> 0</td></tr><tr><td>Pure tone audiometry with visual reinforcement</td><td>> 1</td></tr><tr><td>Whispered speech</td><td>> 3</td></tr><tr><td>Pure tone (play) audiometry</td><td>> 3-4</td></tr></table>			Method	Applicable for Developmental Age (years)	OtoAcoustic Emissions (OAE)	> 0	Auditory Brainstem Responses (ABR)	> 0	Behavioural observation audiometry	> 0	Pure tone audiometry with visual reinforcement	> 1	Whispered speech	> 3	Pure tone (play) audiometry
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OtoAcoustic Emissions (OAE)	> 0															
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Pure tone audiometry with visual reinforcement	> 1															
Whispered speech	> 3															
Pure tone (play) audiometry	> 3-4															
Hypertension	• Measure blood pressure annually. ²² Comments -For patients with spasticity/contractures, may need to do a wrist or thigh blood pressure measurement. Document type of measurement used.															
►Immunizations	• See Centers for Disease Control Recommended Adult Immunization Schedule: http://www.cdc.gov/mmwr/PDF/wk/mm5901-Immunization.pdf .															
►Obesity	• Measure height and weight annually. ²² Comments -Consider weight on home scale in more familiar setting. -Accommodations for patients unable to stand include using a Lift Team, a wheelchair scale, Hoyer Lift, and/or hospital bed which includes a scale.															
►Osteoporosis	• Consider bone mineral density (BMD) screening earlier and at regular intervals for high-risk patients. • Check serum vitamin D 25 OH levels at regular intervals.															
	--	• Although the age to begin screening is unclear, some authors suggest age 40 for patients residing in institutions and age 45 for patients residing in the community. • Multiple sources recommend BMD screening beginning for the general population at age 65 every three to five years if normal baseline test; at age 60 every one to two years if high risk. (AAFP, USPSTF, AACE). ¹⁰														
	Comments -High risk factors in patients with developmental disabilities include: mobility impairments, long term use of antiepileptic drugs or antipsychotics, Down syndrome, Cerebral Palsy, and Prader-Willi syndrome. -High risk factors in the general population include: osteopenia on plain films, history of vertebral fractures, early menopause, chronic steroid use, low body weight, cigarette use and positive family history of osteoporosis. -See FRAX: WHO Fracture Risk Assessment Tool: www.shef.ac.uk/FRAX/ . Note that mobility is not calculated in this assessment tool.															
Prostate Cancer (Men)	--	• Insufficient evidence to recommend routine screening in men under age 75. ²⁰	• Screening not recommended for men over age 75. ²⁰													
	Comments -Family history is often difficult to obtain with this population. -Patients at high-risk include positive family history at an early age and African American men.															

HEALTH PROBLEM	19 – 40 YEARS	40 – 65 YEARS	65 YEARS AND OLDER
Testicular Cancer (Men)	<ul style="list-style-type: none">• Routine screening not recommended.• Prompt assessment and evaluation of testicular problems when young men present with signs and symptoms of testicular disease.²⁰	--	
	Comments <i>-Clinical exam is especially important in this population who may not be able to report symptoms and may have difficulty with the self exam technique.</i>		
Thyroid Disease	<ul style="list-style-type: none">• Perform thyroid stimulating hormone (TSH) test every three years.¹⁶• Thyroid function tests should be performed annually for patients with Down syndrome.¹⁷	<ul style="list-style-type: none">• Perform TSH test annually.	
	Comments <i>-Symptoms of thyroid disease are often not elicited due to cognitive impairment and/or communication difficulties in patients with developmental disabilities.</i> <i>-Consider TSH testing if unexplained change in behavior or level of functioning.</i> <i>-Increased risk for thyroid disease seen in patients with Down syndrome and the elderly.</i>		
Tuberculosis	Screen routinely based on likelihood of exposure. (CDC 2005, AAFP 2008).		
	Comments <i>-Consider PPD skin testing every one to two years for patients who live or work in aggregate settings (board and care homes, intermediate care facilities, day programs).</i>		
Vision	<ul style="list-style-type: none">• Screen annually subjectively or objectively with office-based tests.• Refer to ophthalmology for exam and glaucoma screening at least once before age 40 and by age 30 for patients with Down syndrome.¹⁷	<ul style="list-style-type: none">• Refer for ophthalmologic exam and glaucoma screening every two to three years or as recommended by ophthalmologist.	
	Comments <i>-Screen more frequently for persons with diabetes, those on long-term psychiatric medication, and those with syndromes associated with vision deficits/ocular abnormalities, such as Cornelia de Lange, Fragile X, Down, Smith-Magenis, Tuberous Sclerosis, and Velocardiofacial.¹⁶</i>		

COUNSELING	19 – 40 YEARS	40 – 65 YEARS	65 YEARS AND OLDER
Lifestyle Modification/ Healthy Quality of Life	Discuss: <ul style="list-style-type: none"> • Adequate calcium and vitamin D supplementation. • Advanced directive. • Dental hygiene. • Fall risk assessment and prevention. • Nutrition and physical activity. • Tobacco and substance abuse cessation. • Sexual health, including: contraception, sexually transmitted disease prevention, and healthy relationships. Comments -Include caregivers, health advocates, and parents/family members to help reinforce teaching concepts.		
Medication Review	<ul style="list-style-type: none"> • Review medications at regular intervals with patients and caregivers to assure adherence with regimen and evaluate for side effects and drug interactions. Comments -High rates of polypharmacy exist.		
Safety	<ul style="list-style-type: none"> • Review safety practices per individual circumstance, such as stranger and street safety for patients who live independently; prevention of head trauma in patients with frequent seizures; and street safety for patients with unpredictable behavior. 		

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