


[HOME](#) • [LOGIN](#) • [MY PREFERENCES](#)
[ABOUT NDSS](#)
[RESEARCH](#)
[INFORMATION & RESOURCES](#)
[ADVOCACY CENTER](#)
[NEWS & EVENTS](#)
[COMMUNITY](#)
[General Info](#)
[Health](#)
[Development](#)
[Education & Schooling](#)
[Life Planning](#)
[Find Res](#)

[NDSS Home](#) > [Information & Resources](#) > [Health](#) > [Health Care Guidelines](#)
fo...

Health Topics

- [Alzheimer's Disease](#)
- [Associated Conditions](#)
- [Atlantoaxial Instability](#)
- [Blood Diseases](#)
- [Cosmetic Surgery](#)
- [Dental Care](#)
- [Dental-Periodontal Disease](#)
- [Dimethyl Sulfoxide](#)
- [DS Clinics and UCEDDs](#)
- [Dual Diagnosis](#)
- [Ear, Nose, Throat](#)
- [Early Intervention](#)
- [Endocrine](#)
- [Growth Charts](#)
- [Healthcare Guidelines](#)
- [Healthcare Guidelines - How to Use Them](#)
- [Heart](#)
- [Interferon and Down Syndrome](#)
- [Neurology](#)
- [Physical Therapy](#)
- [Piracetam](#)
- [Sexuality](#)
- [Speech](#)
- [Thyroid](#)
- [Vision & Hearing](#)
- [Vitamin Related Therapies](#)
- [Your Baby and DS](#)

Health Programs

Health Care Guidelines for Individuals with Down Syndrome

Introduction

Individuals with Down syndrome (DS) need the usual health care screening procedures recommended for the general population. For example, children with DS need the usual immunizations and well child care procedures as recommended by the American Academy of Pediatrics.² Immunization practices are continually evolving: be certain to use the most up-to-date protocols.³ Similarly, adults with DS should have health evaluations using the standard accepted practices. However, children with DS have an increased risk of having certain congenital anomalies. Both children and adults may develop certain medical problems that occur in much higher frequency in individuals with DS. Described below is a checklist of additional tests and evaluations recommended for children and adults with DS. These recommendations should take into consideration available local expertise and referral patterns. They are based on our present level of knowledge and should be modified as new information becomes available. Modern primary health care includes educational and developmental concerns within its domain, and therefore we have included information and recommendations specific to these needs of individuals with DS.

These recommendations are a thoughtful composite of the input of many clinicians involved in the care of people with DS. They reflect current standards and practices of health care in the United States of America. They have been designed for a wide audience: for health care professionals who are providing primary care, such as pediatricians, family physicians, internists, and geneticists, as well as specialists, nursing personnel and other allied health professionals, such as physical and occupational therapists, speech-language pathologists, and audiologists. In addition to educators and early intervention providers, these guidelines are designed for parents and other caregivers to use with the professionals who participate in the care of the individual with DS.

Certain recommendations are clearly supported by current



- Changing Lives Partners
 - Changing Lives
-
- 

scientific knowledge. This is the case for the recommendations to look for the presence of congenital heart disease, which occurs in some 50% of infants with DS. In other cases, the recommendations represent our educated guesses. Recognizing the increased frequency of thyroid dysfunction in children with DS, we continue to recommend yearly screening for hypothyroidism. However, we are uncertain as to the appropriate periodicity and nature of the screening: how often, and what constitutes an adequate screening. This question, and others, will be answered by the anticipated development of a large-scale clinical database.

Be certain to use the specific DS growth charts in addition to regular charts to record height and weight (for children from birth to 18 years of age), and head circumference (for children birth to 36 months of age).⁴ If a child is below the third percentile, or if falling off the expected percentiles, consider congenital heart disease, endocrine disorders (thyroid or pituitary), or nutritional factors. Because children with DS have a tendency to become overweight, always use the "Weight v. Height" plots on the growth charts for typically developing children; this will give a more realistic picture of appropriateness of a child's weight.

[Note: Immediately following the recommendations by age, you will find explanations for the specific medical recommendations listed below, descriptive information about other areas of interest to individuals interested in the needs of individuals with Down syndrome, and an updated bibliography.]

About these Health Care Guidelines (Preventive Medical Check List)

These health guidelines continue the series begun in 1981, by Dr. Mary Coleman, and published in Down Syndrome Papers and Abstracts for Professionals (DSPAP), the predecessor of this Down Syndrome Quarterly. The 1992 version was prepared by the members of the Ohio/Western Pennsylvania Down Syndrome Network and published in DSPAP (Volume 15, Number 3, 1992, 1-9) and was based on the 1989 version prepared by Dr. Nancy Roizen, University of Chicago. The 1996 version was the first one prepared by the Down Syndrome Medical Interest Group (DSMIG).

In July, 1994, the Committee on Genetics of the American Academy of Pediatrics published "Health Guidelines for Children with Down syndrome."⁵ Members of DSMIG have been fortunate to work with the Committee during their recent review of their "Health Guidelines" for the purpose of coordinating these efforts and removing differences and incongruities. This version therefore reflects the shared screening protocol, which the Committee will publish in Pediatrics in early 2000. The editor wishes to express his appreciation to Drs. Marilyn Bull and Nancy Roizen for their liaison work with the Committee, and to Dr. Franklin

Desposito, committee chair, and Dr. Tracy Trotter and the other members of the committee for their substantial assistance in this joint effort. The preparation of this revision has been a cooperative effort. The editor has been particularly fortunate to have the expertise of several members of Down Syndrome Medical Interest Group (DSMIG).⁶

This is one of many such compilations. Please see the References, Section C, for a selected list of other protocols.

A NOTE ABOUT FLOW CHARTS: These "Health Guidelines" were prepared with the goal of providing both depth and breadth to the topic of health promotion for individuals with Down syndrome. We trust that this will serve as a reference for families, educators, agencies, and, of course, health care providers. Nevertheless, we recognize the ease and simplicity of using a summary of these guidelines in a one-page graphic format. Such a summary can be placed in the front of a family's medical record book, and likewise, in the front of a medical chart for rapid consultation.

We have included with this document a Flow Chart adapted from Dr. David Smith's document, used at the DS Clinic of Wisconsin in Milwaukee. (See Appendix I, <http://www.denison.edu/collaborations/dsq/recordsheet.html>) There are also two recordsheets, created by Dr. Len Leshin, that can be printed on one page and used for a medical file:

Record Sheet: Birth to Age 12:
<http://www.ds-health.com/recordsheet1.htm>

Record Sheet: Age 13 to Adult:
<http://www.ds-health.com/recordsheet2.htm>

Dr. Brian Chicoine has prepared a variety of materials for providing health care to adults with Down syndrome. These include history questionnaires, review of systems checklists, physical examination forms and an assessment/plan form which includes screening information. You can contact him at the Adult Down Syndrome Clinic at Lutheran General Hospital, Park Ridge, IL at 847-795-2303 if you wish to obtain this material.

What's New

- Additional thyroid screening at 6 months of age.
- Hearing evaluations at birth, and every 6 months thereafter until 3 years.
- Eye evaluations by 6 months of age and yearly.
- Celiac disease screening between 2 and 3 years of age.
- Radiographic screening for atlanto-axial instability once between 3 and 5 years, and then as needed for Special Olympics

Neonatal (Birth to one month)

History: Review parental concerns. Was there a prenatal diagnosis of DS? With vomiting or absence of stools, check for gastrointestinal tract blockage (duodenal web or atresia, or Hirschsprung disease); review feeding history to ensure adequate caloric intake; any concerns about hearing or vision? Inquire about family support.

Exam: Pay special attention to cardiac examination; cataracts (refer immediately to an ophthalmologist if the red reflex is not seen); otitis media; subjective assessment of hearing; and fontanelles (widely open posterior fontanelle may signify hypothyroidism). Exam for plethora, thrombocytopenia.

Lab and Consults: Chromosomal karyotype; genetic counseling; hematocrit or complete blood count to investigate plethora (polycythemia) or thrombo-cytopenia (possible myeloproliferative disorders); thyroid function test-check on results of state-mandated screening; evaluation by a pediatric cardiologist including echocardiogram (even in the absence of a murmur); reinforce the need for subacute bacterial endocarditis (SBE) prophylaxis in susceptible children with cardiac disease; refer for auditory brainstem response (ABR) or otoacoustic emission (OAE) test to assess congenital sensorineural hearing at birth or by 3 months of age. Refer for a pediatric ophthalmological evaluation by six months of age for screening purposes. Refer immediately if there are any indications of nystagmus, strabismus or poor vision. If feeding difficulties are noted, consultation with feeding specialist (occupational therapist or lactation nurse) is advised.

Developmental: Discuss value of Early Intervention (infant stimulation) and refer for enrollment in local program. Parents at this stage often ask for predictions of their child's abilities: "Can you tell how severe it is?" This is an opportunity to discuss the unfolding nature of their child's development, the importance of developmental programming, and our expectation of being able to answer that question closer to two years of age.

Recommendations: Referral to local DS parent group for family support, as indicated.

Infancy (1 - 12 months)

History: Review parental concerns. Question about respiratory infections (especially otitis media); for constipation, use aggressive dietary management and consider Hirschsprung disease if resistant to dietary changes and stool softeners. Solicit parental concerns regarding vision and hearing.

Exam: General neurological, neuromotor, and musculoskeletal examination; must visualize tympanic membranes or refer to ear, nose and throat (ENT) specialist,

especially if suspicious of otitis media.

Lab and Consults: Evaluation by a pediatric cardiologist including echocardiogram (if not done in newborn period); remember to consider progressive pulmonary hypertension in DS patients with a VSD or atrioventricular septal defect who are having little or no symptoms of heart failure in this age group. Auditory brainstem response test (ABR) by 3 months of age if not performed previously or if previous results are suspicious. Pediatric ophthalmology evaluation by six months of age (earlier if nystagmus, strabismus or indications of poor vision are present). Thyroid function test (TSH and T4), at 6 and 12 months of age. Evaluation by ENT specialist for recurrent otitis media as needed.

Developmental: Discuss early intervention and refer for enrollment in local program (if not done during the neonatal period). This usually includes physical and occupational therapy evaluations and a developmental assessment.

Recommendations: Application for Supplemental Security Income (SSI) (depending on family income); consider estate planning and custody arrangements; continue family support; continue SBE prophylaxis for children with cardiac defects.

Childhood (1 year to 12 years)

History: Review parental concerns; current level of functioning; review current programming (early intervention, preschool, school); ear problems; sleep problems (snoring or restless sleep might indicate obstructive sleep apnea); constipation; review audiologic and thyroid function tests; review ophthalmologic and dental care. Monitor for behavior problems.

Exam: General pediatric and neurological exam including evaluation for signs of spinal cord compression: deep tendon reflexes, gait, Babinski sign. Include a brief vulvar exam for girls. Use Down syndrome growth charts, as well as growth charts for typically developing children. Be sure to plot height for weight on the latter chart.

Lab and Consults: Echocardiogram by a pediatric cardiologist if not done previously; Thyroid function test (TSH and T4) yearly; behavioral auditory testing every 6 months until 3 years of age, then yearly. Continue regular eye exams every year if normal, or more frequently as indicated. Between 3 years and 5 years of age, lateral cervical spine x-rays (neutral view, flexion and extension) to rule out atlanto-axial instability: have the radiologist measure the atlanto-dens distance and the neural canal width. X-rays should be performed at an institution accustomed to taking and reading these x-rays. Initial dental evaluation at two years of age with follow-ups every six months. At 2-3 years of age, screen for celiac disease with IgA antiendomysium antibodies, as well as total IgA. Administer Pneumococcal vaccine at 2 years of age.

Developmental: Enrollment in appropriate developmental or educational program; complete educational assessment yearly, as part of Individualized Family Service Plan (IFSP) for children from birth to 3 years of age, or Individualized Educational Plan (IEP) from age four until the end of formal schooling. Evaluation by a speech and language pathologist is strongly recommended to maximize language development and verbal communication. An individual with significant communication deficits may be a candidate for an augmentive communication device.

Recommendations: Twice daily teeth brushing. Total caloric intake should be below recommended daily allowance (RDA) for children of similar height and age. Monitor for well-balanced, high fiber diet. Regular exercise and recreational programs should be established early. Continue speech therapy and physical therapy as needed. Continue SBE prophylaxis for children with cardiac defects. Monitor the family's need for respite care, supportive counseling and behavior management techniques. Reinforce the importance of good self-care skills (grooming, dressing, and money handling skills).

Adolescence (12 to 18 years)

History: Review interval medical history, questioning specifically about the possibility of obstructive airway disease and sleep apnea; check sensory functioning (vision and hearing); assess for behavioral problems; address sexuality issues.

Exam: General physical and neurological examination (with reference to atlanto-axial dislocation). Monitor for obesity by plotting height for weight on the growth charts for typical children. Pelvic exam if sexually active, only. (See Consults, below.) Perform a careful cardiac exam in adolescents, looking for evidence of valvular disease. Lab and consults: Thyroid function testing (TSH and T4) yearly. Hearing and vision evaluations every year. Repeat screening cervical spine x-rays as needed for Special Olympic participation. Echocardiogram if evidence of valvular disease on clinical exam. Consult with Adolescent Medicine practitioner or a gynecologist experienced in working with individuals with developmental disabilities to address issues of sexuality and/or for pelvic examination for sexually active teenager. Continue twice-yearly dental exams.

Developmental: Repeat psycho-educational evaluations every two years as part of Individualized Educational Plan (IEP). Monitor independent functioning. Continue speech/language therapy as needed. Health and sex education, including counseling regarding abuse prevention. Smoking, drug, and alcohol education.

Recommendations: Begin functional transition planning (age 16). Consider enrollment for SSI depending on family income. SBE prophylaxis needed for individuals with cardiac disease.

Continue dietary and exercise recommendations (see childhood, above). Update estate planning and custody arrangements. Encourage social and recreational programs with friends. Register for voting and selective service at age 18. Discuss plans for alternative long term living arrangements such as community living arrangements (CLA). Reinforce the importance of good self-care skills (grooming, dressing, and money handling skills).

Adults (over 18 years)

History: Interval medical history. Ask about sleep apnea symptoms. Monitor for loss of independence in living skills, behavioral changes and/or mental health problems. Symptoms of dementia (decline in function, memory loss, ataxia, seizures and incontinence of urine and/or stool). This may also represent spinal cord compression from atlanto-axial subluxation.

Exam: General physical and neurological examination (with reference to atlanto-axial dislocation). Monitor for obesity by plotting height for weight. Cardiac exam: listen for evidence of mitral valve prolapse and aortic regurgitation: confirm suspicions with echocardiogram. Sexually active women will need Pap smears every 1-3 years following the age of first intercourse. For women who are not sexually active, single-finger bimanual examination with finger-directed cytology exam. Screening pelvic ultrasound every 2-3 years for women who refuse or have inadequate follow-up bimanual examinations. This may require referral to an Adolescent Medicine practitioner or a gynecologist with experience with individuals with special needs. Otherwise, pelvic ultrasound may be considered in place of pelvic examinations. Breast exam yearly by physician.

Lab and consults: Annual thyroid screening (TSH and T4). Ophthalmologic evaluation every two years (looking especially for keratoconus and cataracts). Repeat cervical spine x-rays as needed for Special Olympic participation. Continue auditory testing every two years. There are two different suggestions for mammography: Dr. Heaton recommends yearly study after age 50; begin at age 40 for women with a first-degree relative with breast cancer. Dr. Chicoine suggests a mammogram every other year beginning at 40, and yearly beginning at 50. Continue twice-yearly dental visits. Mental health referral for individuals with emotional and behavioral changes.

Developmental: Continue speech and language therapy, as indicated. For individuals with poor expressive language skills, consider referral for augmentive communication device. Discuss plans for further programming/vocational opportunities at age 21 or when formal schooling ends. Be aware that accelerated aging may affect functional abilities of adults with DS, more so than Alzheimer disease.

Recommendations: Discuss plans for alternative long term

living arrangements such as community living arrangements (CLA). SBE prophylaxis needed for individuals with cardiac disease. Continue dietary and exercise recommendations (see childhood, above). Update estate planning and custody arrangements. Encourage social and recreational programs with friends. Register for voting and selective service at age 18. Reinforce the importance of good self-care skills (grooming, dressing, and money handling skills). Bereavement counseling for individuals who have experienced the loss of an important person in their life, either via death or by other circumstances: sibling moves away after marriage, or goes off to college.

The following are an elaboration of the recommendations made above, as well as other information designed to promote optimal health care for individuals with Down syndrome:

Cardiac: Congenital heart disease is reported to occur in 30 - 60% of children with DS. Ventricular septal defects and complete atrioventricular septal defects are among the most common. A serious cardiac defect may be present in the absence of a murmur because of the increased tendency of children with DS to develop early increases in pulmonary vascular resistance which reduces the left to right intracardiac shunt, minimizes the heart murmur, and prevents symptoms of heart failure and respiratory problems. Children with DS with a significant cardiac defect who seem to be doing clinically well or getting better, especially during the first 8 months of life, may be developing serious pulmonary vascular changes. Timely surgery, frequently during the first 6 months of life, may be necessary to prevent serious complications. Therefore, all infants and children need to have an evaluation by a pediatric cardiologist, preferably before three months of age, which should include an echocardiogram. In some tertiary care centers, an echocardiogram alone is satisfactory when it will be evaluated by a pediatric cardiologist. If this is not available, a full evaluation by a pediatric cardiologist is mandatory. For the older child, who has never had a cardiac evaluation and who has no signs of cardiac disease, a screening echo-cardiogram is recommended. Adolescents and young adults with no known intracardiac disease can develop valve dysfunction and should be evaluated by clinical examination at age 18, especially prior to dental or surgical procedures. [See References, Section G, Geggel RL, et al.] There is a 57% incidence of mitral valve prolapse and approximately a 10% risk of aortic regurgitation. The finding of a click or murmur should be followed by an echocardiogram. Susceptible individuals will need SBE prophylaxis.

Dental Care: The orofacial features of individuals with DS contribute to a variety of potential problems in regards to dental care. For example, the eruption of teeth is usually delayed and often occurs in an unusual order. Primary and permanent teeth may be missing. Small or misshapen teeth are found, and severe crowding can occur because of the

small oral cavity. Orthodontic treatment may be necessary. Mouth breathing, related to the small nasal airway, contributes to fissured tongue and lips. Periodontal disease can occur as early as the teen years, and routine brushing combined with dental visits every 6 months play a key role in preventing tooth loss.

Ears/Audiology: Hearing loss is a significant area of concern for individuals with DS. Infants and children may have a sensorineural loss, a conductive loss (related to middle ear effusions) or both. All infants with DS should have an objective measure of hearing performed at birth, if possible, or within the first 3 months of life. The most common method in widespread use is the measurement of auditory brainstem responses (ABR), also known as brainstem auditory evoked response (BAER). Two screening methods include ABR screening in the newborn nursery, and evoked oto-acoustic emission testing. The typical behavioral audiology requires a developmental age of 7-8 months. Consequently, all children with DS need an objective measure when tested in the first 12 months. Subsequently, behavioral audiology may be appropriate. Most children with DS have very small ear canals, making it difficult to examine them properly with the instruments found in the pediatrician's office. Consequently, it may be necessary to refer the child to an Ear, Nose, and Throat physician to visualize the tympanic membranes using the microscopic otoscope. An ENT physician should evaluate all children with an abnormal hearing evaluation and/or tympanogram in order to aggressively manage treatable causes of hearing loss (using antibiotics and/or tympanostomy tubes as indicated). Fluid can accumulate as early as the neonatal period and aggressive otologic care can minimize the effect of any hearing loss on language development.

Individuals with Down syndrome may begin to develop hearing loss in their second decade, which, if undetected may lead to behavioral symptoms which could be misinterpreted as a psychiatric disorder.

ENT: The midfacial hypoplasia (under-development) that is characteristic of individuals with DS leads to increased difficulty with narrow airways. The narrow nares (nostrils) manifest themselves as noisy breathing in the infant; narrow openings to paranasal sinuses predisposes children to frequent sinusitis/naso-pharyngitis. These infections, manifested by purulent nasal discharge, should be aggressively treated. The narrow trachea can result in recurrent croup. In addition, infants with DS have an increased likelihood of tracheomalacia (partial collapse of the trachea).

Obstructive airway disease has been recognized as a significant problem for children and adults with DS. Symptoms include snoring, unusual sleeping positions (sitting up or bending forward at the waist with head on knees), fatigability during the day, reappearance of napping in older

children or behavior change. Individuals with these symptoms should be evaluated completely via detailed history (looking specifically for evidence of sleep apnea), physical examination with regard to tonsillar size, and prompt referral to an ENT physician for further evaluation (eg. assessment of adenoidal size). In a number of children, hypotonicity and collapse of the airway leads to similar symptoms in the absence of obstruction caused by lymphoid tissue. Surgical intervention may be necessary to avoid hypoxemia and possible cor pulmonale (right heart failure). In the absence of surgically correctable problems, supplemental O₂ therapy under pressure (such as bi-pap) may be indicated.

A recent study from Israel notes that children with DS have significant sleep fragmentation with frequent awakenings and arousals that are not related to obstructive sleep apnea syndrome. [See References, Section K, Levanon et al.]

Two studies point to the importance of keeping children with DS in the hospital overnight following tonsillectomy and adenoidectomy, because of higher rate of postoperative respiratory complications. [See References, Section K, Bower et al, and Goldstein et al.]

Endocrine: The incidence of thyroid disease is significantly increased among individuals with DS of all ages. Normal thyroid hormone levels are necessary for growth and cognitive functioning. The signs of hypothyroidism may be subtle in individuals with DS and may be attributed to the DS itself. Therefore, screening is recommended on a yearly basis by monitoring TSH and T₄ levels. Since autoimmune conditions are common in individuals with DS, evaluation of suspected hypothyroidism in the school age child should include thyroid antibodies to look for thyroiditis. Some infants and young children have a condition known as idiopathic hyperthyrotropinemia, with borderline abnormal TSH with normal T₄. This may reflect a neuroregulatory defect of TSH, which, when studied by 24 hour sampling, varies between normal levels and very high levels. Therefore, some centers recommend repeating the TSH and T₄ every six months, withholding treatment unless the T₄ is low. Immune-mediated hyperthyroidism also occurs in children and adults with DS. High sensitivity TSH levels will be abnormally low in these cases. In addition, weight loss, GI symptoms and heat intolerance are often seen.

Diabetes mellitus, recognized to be an autoimmune condition, also occurs more frequently in individuals with with DS, with a prevalence rate between 1.4 and 10.6%. [References, Section M, Anwar et al.]

There has been some discussion about the use of human growth hormone in children with DS in response to a report that suggested that children with DS have an abnormality of growth hormone secretion. This issue has been addressed by members of the Down Syndrome Medical Interest Group, and

published in Down Syndrome Quarterly, Vol 1, Number 1 (March, 1996), page 8: "On the basis of the available evidence, and until the recommended scientific studies are completed, the uncontrolled use of hormonal treatments such as growth hormone in children with Down syndrome is not supported by the Down Syndrome Medical Interest Group." A recent study from Sweden and Australia reveals that treatment with human growth hormone did result in "normal height velocity but did not affect head circumference or mental or gross motor development." [See References, Section M, Anneren G, Tuvemo T, Sava VR et al.]

Feeding/Nutrition: Infants with DS may initially experience difficulty with coordination of suck and swallow. They may need assistance of a feeding specialist (such as nurse specialists, occupational therapists, speech-language pathologists) or lactation specialists, if the mother is nursing. Later on, toddlers may have difficulty with texture progression. Significant disruption of family activities, involvement of more than two professionals indicates that a consultation with a multi-disciplinary feeding team may be warranted. [See References Section N, Medlen.] Remember that there is up to a 10% difference in basal metabolic rate for individuals with DS.

Follow % Ideal Body Weight, calculated as follows: plot child's height on growth chart for typically developing children and determine the age for which this height is at the 50th percentile. Determine the weight at the 50th percentile for this height. Divide the actual weight of the child by this determined weight and multiple by 100. Goal is 90% - 100% IBW. Chart this sequentially.

Hematology: Leukemia is more common in children with DS than in the general population. Nevertheless, this is rare. Most leukemia in children less than 3 years of age is non-lymphocytic leukemia (usually acute myelogenous leukemia). Children with DS usually respond very favorably to standard treatment, going into remission easily. In the newborn period, there is a 10% incidence of myelo-proliferative disorder ("leukemoid reaction") which in some instances develops into acute megakaryoblastic leukemia. Polycythemia has been frequently observed in the newborn period; in one series, as high as 64% of children studied were affected

Infectious Disease/Immunology: Persons with DS who have serious recurrent respiratory and systemic infections are often evaluated for immune function. Consider measuring the IgG subclasses in such individuals. Total IgG level may not disclose any abnormality, although there may be a deficiency of IgG subclasses 2 and 4 and an increase of IgG subclasses 1 and 3. There is a significant correlation between the decreased IgG subclass 4 levels and bacterial infections. The mechanism is not known but theories include the possibility that this subclass plays a role in pulmonary host defense or possibly a deficiency of selenium. Intravenous gamma globulin replacement therapy should be a consideration in a

person with DS who presents with serious recurrent bacterial infections and documented IgG subclass 4 deficiency.

The cellular immunity deficits described in individuals with DS have the greatest documented clinical impact on gingivitis and periodontal disease.

Children with chronic cardiac and respiratory disease are candidates for use of pneumococcal, respiratory syncytial virus, and influenza vaccines.

Eye/Vision: Congenital cataracts are a serious problem for infants with DS, leading to vision loss if not detected and treated. The absence of a red reflex is sufficient cause for immediate referral to a pediatric ophthalmologist, as are strabismus and nystagmus. Routine evaluations should begin at 6 months of age, and be performed annually thereafter. Refractive errors are common and will be detected during these evaluations, as would serious, but rarer, conditions, such as keratoconus. Stenotic nasolacrimal ducts may lead to tearing in infancy. Blepharitis and conjunctivitis occur frequently. Keratoconus occurs more frequently in adolescents with DS than in the typical child.

Orthopedic Disorders & Atlantoaxial Instability (AAI): Ligamentous laxity is responsible for a number of orthopedic difficulties in individuals with DS. Interestingly, congenital hip dislocation is not commonly encountered. Hip dislocation is more often seen in the older child and the adolescent. Chronic patellar dislocation can lead to gait disturbances in the adolescent. Atlantoaxial instability is a term used to describe increased mobility