



Down Syndrome Association
of West Michigan *Possibility. Promise. Potential.*

Guide for Parents of Teens and Young Adults with Down Syndrome

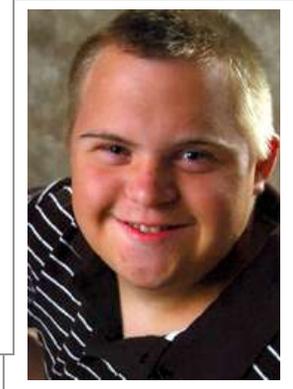
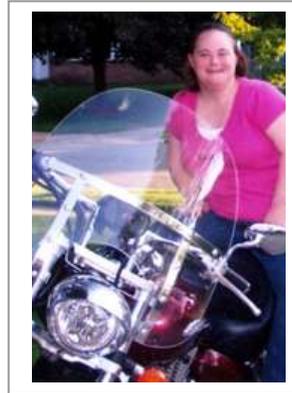


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Dear Reader,

Thank you for requesting information on Down syndrome.

The Down Syndrome Association of West Michigan (DSAWM) is a nonprofit organization committed to increasing opportunities for individuals with Down syndrome. The mission of the Down Syndrome Association of West Michigan is to be a resource and advocacy organization, promoting public awareness and supporting lifelong opportunities for individuals with Down syndrome and their families.

The DSAWM is proud to offer services to our ever-growing population of teens and young adults with Down syndrome. At the forefront of these services is THREADS, our group supporting teens and young adults with Down syndrome and their families as they navigate the transition process from school to adult life. Families with teenagers age 14 and over are invited to explore this process with us. The focus of THREADS is to explore opportunities by learning from, and sharing experiences with, other parents. Activities for teens and young adults are scheduled during the bi-monthly parent meetings, and additional activities occur monthly.

- The DSAWM also holds annual conferences and periodic workshops. Conferences and workshops are open to anyone interested in Down syndrome - family members, caregivers and involved professionals. We strive to have professionals speak to us about relevant medical, educational and transition issues. The THREADS bi-monthly parent meetings also offer a forum for parents to meet on an informal basis to ask questions and learn from each other.
- The DSAWM is proud to offer an extensive lending library of books, periodicals and videos on relevant topics for use by members and professionals.
- Annually, we hold a Buddy Walk to promote public awareness and acceptance of individuals with Down syndrome.
- The Association participates in community and health expos and legislative forums.
- The DSAWM is involved in many public awareness activities including book distributions to area schools and universities and speaking engagements. We have been the subject of several local radio and TV news stories and have been featured in many newspaper articles, all to disseminate accurate information about Down syndrome and to enhance the public's opinion of individuals with Down syndrome.

To contact the Down Syndrome Association of West Michigan, please call 616-956-3488 or e-mail info@dsawm.org.

Please do not hesitate to call. We look forward to hearing from you!

Promoting Health in Adults with Down Syndrome

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Health is more than the absence of disease. It also involves a lifestyle that promotes physical, mental and spiritual well-being. This is as true for adults with Down syndrome as it is in the general population. This paper focuses on the physical and mental health and then discusses the interaction of these two aspects in of adults with Down syndrome.

A HEALTHY LIFESTYLE

To maximize physical health requires a multi-faceted approach: living a healthy lifestyle, following certain preventive measures, participating in appropriate health screening and treating problems early in their course. A healthy lifestyle includes an appropriate diet, regular exercise, adequate sleep and opportunities for social and creative use of one's time.

Diet

Although coronary artery disease (heart attacks) and hypertension (high blood pressure) are less common in adults with Down syndrome, the dietary and exercise recommendations for people with Down syndrome are similar to those for the general population. These include following a diet like the one described in the "pyramid" diet program which recommends limiting fat intake and encourages a diet high in complex carbohydrates.

Obesity is more common in persons with Down syndrome. A recent study found that children with Down syndrome have a basal metabolic rate that is lower than that seen in the general population. On average, at rest, those with Down syndrome burned 200-300 less calories per day. Reduction in calories alone led to nutrient deficiencies and, therefore, it is necessary to burn 200-300 calories more per day through activity to prevent weight gain. In addition, when making dietary recommendations to prevent weight gain or promote weight loss, we have had better success with offering choices such as fresh fruit or vegetables and low fat foods such as popcorn or pretzels rather than offering no dessert or snacks. Just as in the general population, better success is achieved in the long run by following good general nutrition principles and regularly exercising than is by dieting.

Exercise

Twenty to thirty minutes of aerobic exercise at least three days a week is recommended. Aerobic exercise includes such activities as walking, running, swimming, biking and cross country skiing. Prior to starting an exercise program, a physical exam and health screening are indicated. Forty to fifty percent of babies born with Down syndrome have congenital heart

disease. Some adults with Down syndrome have had these surgically corrected and all they need is antibiotics when they go to the dentist (see below). Others have not had the heart condition corrected and many will need ongoing treatment. A few studies indicate that some adults with Down syndrome develop disease of their heart valves later in life. Exercise restriction may be necessary depending on the type of congenital or acquired problem. Physical examination and an echocardiogram and/or a stress test (treadmill test) may be necessary to determine the safety of exercise.

Another health problem that may make modification in exercise necessary is atlanto-axial instability. This condition in which the first vertebrae (bone) in the neck slips on the second is more common in persons with Down syndrome. The exact significance is not always clear but it may make contact sports or sports which could jar the neck more dangerous. For this reason, the Special Olympics requires lateral neck x-rays prior to participation. The need for these x-rays, however, is the subject of ongoing research and debate.

Exercise is believed to have many benefits. Although reduction in coronary artery disease and hypertension would not appear to be major benefits in adults with Down syndrome (because of the infrequent occurrence of these diseases), exercise probably has several other benefits. Although this is not as well studied in persons with Down syndrome, in the general population exercise has been shown to help improve overall sense of well-being, self-esteem, and also the overall fitness of those who regularly participate. It is an excellent mechanism to help people deal with stress and reduce its effects.

Sleep

Adequate sleep is also an important part of a healthy lifestyle. The exact number of hours of sleep needed varies from person to person, but enough sleep to feel energetic throughout the day is recommended. Many families have commented that their son or daughter sleeps bent at the waist with his or her head resting on the legs. While this does not seem to be a significant problem or necessarily indicate a significant underlying problem, there are some sleep problems that can occur. Sleep apnea may be more common in persons with Down syndrome. Sleep apnea is often associated with restless sleeping, irregular breathing pattern, snoring or snorting sounds and daytime somnolence. It is generally caused by obstruction of the airway in persons with Down syndrome and may cause heart disease and several other health problems.

Social Opportunities

We have demonstrated that adults with Down syndrome who have social opportunities tend to be healthier. Meeting with friends, being with family, and having opportunities for creative activities at work and socially all contribute to our sense of self esteem, well-being and happiness. Studies in the general population and our study in persons with Down syndrome support the idea that these contribute to the health of an individual. In the general population, depression and stress have been shown to be associated with decreased immunity and increased physical illness.

PREVENTATIVE MEASURES

Immunizations

In addition to the preventive measures discussed as part of a healthy lifestyle, there are several other recommendations to promote health. Immunizations are recommended for all adults. After receiving the usual immunizations of childhood, a diphtheria-tetanus booster is recommended every ten years. For persons older than sixty five and for persons with certain

chronic illnesses, an annual influenza (flu) shot is recommended as well as a pneumonia vaccine every seven years. Some investigators have recommended that adults with Down syndrome should receive the flu and pneumonia vaccines in their fifties instead of waiting until 65 because of their relatively weaker ability to fight infections. Down syndrome is not one of the conditions that require a child or younger adult to receive these immunizations, however, those with associated health problems such as congenital heart disease or recurrent pneumonia should receive them. In addition, flu vaccine is recommended annually for people living in residential facilities.

Hepatitis B immunization is recommended for residents of residential facilities. In addition, some studies have found that other adults with Down syndrome are at greater risk for getting hepatitis B, possibly in workshops or other settings. Although hepatitis B transmission is generally considered to occur through blood (blood transfusions, shared needles) and sexual activity, hepatitis B virus is actually shed in all body secretions. In any setting where regular contact with an infected person's secretions can occur or where hygiene may be limited, hepatitis B may be transmitted. Hepatitis B immunization is given in a three shot series. The second and third doses are given one and six months after the first. A blood test (Hepatitis B surface antibody) should be drawn one to two months after the third dose to confirm the effectiveness of the vaccine in providing immunity.

Antibiotic Prophylaxis

Another preventive measure is antibiotic prophylaxis. Some people with congenital or acquired heart disease need antibiotics, to prevent an infection in their heart, before and after going to the dentist (even for a routine cleaning) or when they undergo procedures to their gastro-intestinal or urinary tracts. This is usually done with Amoxicillin 500 mg tabs: 6 tabs 1 hour before the procedure and 3 tabs 6 hours after. For those allergic to amoxicillin (or other penicillins), alternative regimens are recommended.

Education

Sexuality and reproduction are areas in which adults with Down syndrome often need education. Men with Down syndrome have generally been considered to be sterile, although there is one reported case of a man with Down syndrome fathering a child. Women with Down syndrome may have a slightly reduced fertility rate but can have children. Roughly 50% of children born to a woman with Down syndrome will have Down syndrome. Birth control pills, Depo-Provera, Norplant, and other forms of birth control generally are considered to have fewer complications than tubal ligation. Depo-Provera, an injection of progesterone every 3 months is both effective and allows for easy compliance. Barrier methods such as condoms and diaphragms are often not effective because of limitations in the ability of the person to use it correctly each time. Other than condoms, birth control measures do not prevent AIDS and other sexually transmitted diseases. Also, sexual abuse is a concern and, therefore, education is necessary even if birth control is being used or sterilization has been performed.

HEALTH SCREENING AND TREATMENT OF MEDICAL PROBLEMS

In addition to a healthy lifestyle and preventive measures, appropriate health screening measures are recommended. The Adult Down Syndrome Center of the Advocate Health System was developed in response to a request by the National Association for Down Syndrome (NADS), a local parent group, to fill the need for screening and other health care for adults with Down syndrome. The staff of NADS surveyed its members to determine the needs

of the patients and their families. The staff of the Center reviewed the available literature, especially the Preventive Medicine Checklist developed by the Ohio and Western Pennsylvania Down Syndrome Clinic Directors Group. Based on this available information and the subsequent care of over 500 patients, we recommend the following health screening. An annual health maintenance evaluation including a review of the patient's history and a physical exam will help find problems early in their course and provide an opportunity to review the issues of a healthy lifestyle. Routine health screening that is recommended for the general population is recommended for adults with Down syndrome as well. This would include mammograms, pap smears, screening for colo-rectal cancer and cholesterol screening. With further study these recommendations could change depending on the frequency of these diseases in persons with Down syndrome.

The history and physical exam should give special attention to the areas in which adults with Down syndrome have problems that are more common or present differently than in the general population.

Skin

A number of skin problems are seen frequently in adults with Down syndrome. Dry skin (xeroderma) is very common. The handout provided to our patients is included at the end of this paper. Seborrheic dermatitis occurs frequently also. This is often seen on the scalp as dandruff. Anti-dandruff shampoos such as Head and Shoulders, Denorex and Selsun Blue are helpful. Tar shampoos are also a good treatment. Recent evidence suggests that some of this may be caused by a fungal infection and, therefore, some people benefit from Nizoral shampoo, an anti-fungal prescription shampoo. When it occurs on other parts of the body, a steroid cream can help clear it.

Another skin condition, folliculitis (infection or inflammation of the hair follicles) can be treated by washing with an anti-bacterial soap such as Dial or Lever 2000. Since so many patients also have dry skin, we generally recommend Lever 2000 or Dial with moisturizer which tend not to dry out the skin. Folliculitis is often on the back or buttocks which can be hard to reach. Therefore, a soft brush on a handle is helpful.

Tinea infections (fungal infection of the skin) and onychomycoses (fungal infection of the nails) occur more commonly. Tinea pedis (athlete's foot) is treated with good daily cleaning followed by thorough drying. Since the fungus thrives in warm, moist environments, it is sometimes necessary to change socks in the middle of the day to keep the feet adequately dry. Anti-fungal preparations such as Desenex, Tinactin, or Lotrimin help kill the fungus. The treatment of onychomycoses is problematic. Topical preparations usually don't work well. Until recently, the oral medications (pills) for this problem had to be taken for 6 to 12 months and could cause inflammation of the liver. Recently, Sporanox was approved for use in onychomycoses. It only has to be taken for 3 months and seems to cause less liver problems. It, however, is very expensive. Toenail removal is painful and if the nail is allowed to grow back the fungal infection can recur. If the toenail bed is destroyed so that the nail does not grow back, then the toe is left without its protective nail and can be sensitive to even mild trauma. Since the condition does not usually cause any symptoms, daily foot soaks followed by thorough drying and keeping the toenails cut straight across is a simple but helpful recommendation. Sometimes podiatric care is needed to aid in toenail cutting.

Eyes

Poor visual acuity (eyesight) is a common problem just as it is in the general population. However, the adult with Down syndrome may have a difficult time perceiving the problem or communicating it to someone who can help. If work skills or other daily skills deteriorate, the cause could be as simple as a decline in eyesight.

Cataracts are more common. Keratoconus (thinning of the cornea) is more common and can be a sight-threatening problem that needs regular follow-up by an ophthalmologist. Corneal transplant becomes necessary in the later stages of keratoconus. Strabismus (crossing of the eyes) usually needs to be corrected in childhood for any chance of significant vision in the crossed eye. Blepharitis (inflammation or infection of the eyelids) is treated by careful, regular washing with a warm washcloth and half strength baby shampoo and sometimes antibiotic drops. Good treatment of seborrheic dermatitis of the scalp may also be beneficial because sometimes the blepharitis is related to the scalp condition. The increased frequency of eye problems requires regular screening by an eye doctor, probably at least every two years and more frequently if problems exist.

Hearing/Ears

Hearing loss is also more common in adults with Down syndrome. It can be from reversible causes such as ear wax or fluid behind the drum or it can be permanent and associated with inner ear problems. Some inner ear hearing losses are stable for many years; others are progressive. These inner ear problems are made worse if excessive ear wax or fluid behind the eardrum (in the middle ear) is overlaid on the irreversible inner ear hearing loss. Ear wax (cerumen) building up until it obstructs the canal is common in persons with Down syndrome. Wax softening drops can be used to help the natural body function to remove the wax. However, ear drops should not be used if the eardrum is punctured or perforated. We advise people to “never put anything smaller than your elbow in your ear” because anything, such as cotton swabs, put down the canal interrupts the body’s natural ability to remove wax and may push the wax further inward, thereby making it more difficult to remove. Sometimes the excess wax must be irrigated or suctioned out. Another cause of hearing loss is fluid behind the eardrum and this may require placement of tubes through the eardrum to drain the fluid and increase hearing. A person who seems to be losing daily life skills may have a problem as simple as decreased hearing from wax obstruction. Removal of the wax and restoration of hearing can sometimes help restore skills.

Many adults with Down syndrome have an inner ear problem which affects the ability to hear high pitched sounds, including many speech consonants. Because inner ear hearing loss cannot be seen by an exam of the outer ear and eardrum, a referral for a comprehensive audiologic (hearing) evaluation may be indicated to clarify hearing status and to identify the high frequency (pitch) impairment. This problem can gradually develop and contribute to increased misunderstandings, attention difficulties and general confusion especially in groups or in any situation that is not optimally quiet. Sometimes the best option for hearing loss is a hearing aid. Careful attention to fitting and sometimes a gradual increase in the amount of time the aid is worn will help adults with Down syndrome adjust to the device.

Sinuses

Sinusitis (sinus infection) is usually readily treatable with antibiotics and decongestants. The acute presentation includes fever, headache and face pain. It may present as chronic runny nose or congestion. Sometimes the ability to communicate the pain is lacking and, therefore, a change in behavior is noted that will resolve with appropriate treatment of the sinus condition.

Teeth and Gums

Periodontal (gum) disease is common and can be prevented by good teeth brushing, flossing, and regular dental evaluations. Persons with Down syndrome often need more dental care to keep the gums healthy because of a tendency for problems in this area. Most people who lose their teeth do so because of gum disease. Cavities are actually less common in persons with Down syndrome than in the general population.

Gastrointestinal

Constipation is fairly common but can usually be managed by increasing the fiber in the diet by eating more fruits, vegetables and whole grains. Drinking plenty of water (6-8 eight ounce glasses per day) also helps. Metamucil or other fiber supplements can also be used. Hernias can also occur in adults with Down syndrome and surgical correction is the only cure, although trusses can be used for symptom relief. The need for surgery should be based on the symptoms associated with the hernia, the general activity level, the age, the size of the hernia and the overall health of the person. Another problem, incontinence of stool is not a common finding in our patients although hygiene issues are a problem for some and may need further attention with education.

Gynecologic

General gynecologic care includes daily care issues and evaluation in the physician's office. Education is important for self-care and to prepare the woman for the office evaluation. In the office, a slow, gentle approach is often all that is needed although sometimes light sedation is necessary. Modified exams can be done to get a pap smear and sometimes an ultrasound of the pelvis can be done to provide some information that is not obtained in the exam.

Dysmenorrhea (painful periods) is common in women with Down syndrome just as it is in the general population. It is usually effectively controlled with Tylenol, Advil, Nuprin or similar medications. A woman with Down syndrome may have difficulty communicating her pain and the only outward sign will be a behavior change around the time of her period. Use of medication to reduce cramping can be extremely helpful in reducing the behavior changes. Likewise, behavioral or other changes that occur cyclically before the period should be noted and the woman evaluated for premenstrual syndrome (PMS).

Urinary

Incontinence of urine that starts in adulthood may be more common than in the general population. Urinary tract infections, nervous system disorders, behavioral issues and anatomic problems must all be considered. We have seen a number of adults with urinary retention (inability to adequately empty the bladder) that leads to incontinence and often discomfort. In addition, incontinence can be seen in Alzheimer's disease.

Orthopedic

In addition to atlanto-axial instability being an issue in exercise as previously discussed, it must be considered as part of evaluation prior to any surgery. Persons with Down syndrome who have atlanto-axial instability have received severe neck injuries during surgery when their necks were extended to allow for placing the endotracheal (breathing) tube. The anesthesiologist must make adjustments to prevent this. Even with normal x-rays, special care should be given to the neck of a person with Down syndrome during anesthesia.

Another more common orthopedic problem is hallux valgus (bunions). They can usually be treated conservatively with wide shoes, acetaminophen (Tylenol) or anti-inflammatory agents (Advil, Nuprin, ibuprofen) and soaking in warm water. Surgery is indicated when the pain is not responding to these treatments and is severe enough to warrant the discomfort of the surgical correction.

Neurologic

Alzheimer's disease is a frequently voiced concern about people with Down syndrome. At autopsy, studies have shown plaques in the brains of all adults with Down syndrome over the age of 35. These plaques are similar to the plaques seen in persons with Alzheimer's disease. Neurofibrillary tangles (also seen in Alzheimer's disease) have not been found generally until after the age of 50 and then only in approximately the same percentage as in the general population. Clinical findings and recent studies suggest that the tangles correlate better with the clinical manifestation of Alzheimer's disease and that the percentage of adults with Down syndrome who develop Alzheimer's disease may be similar to the general population. There is a tendency for persons with Down syndrome to develop Alzheimer's disease at a younger age (50's) than in the general population (70's). Most importantly, many other medical and psychological problems that are potentially reversible can mimic Alzheimer's disease and should be checked prior to making the diagnosis. These include poor hearing, poor vision, hypothyroidism, brain tumors, vitamin B-12 deficiency and depression.

MENTAL HEALTH

It is also important to evaluate for mental health problems. However, it helps to understand the aspects of a healthy life that promote mental health prior to understanding mental illness.

The ability to communicate is a strong factor in mental health. For those for whom communication skills are weak, speech therapy, communication boards, sign language and other methods of communication can be helpful. Even when verbal skills are good, the ability to communicate concepts such as frustration and sadness may be limited. Sometimes it is helpful to have opportunities to gather with other adults with developmental disabilities to share experiences. This type of interaction and support can help the individual express his or her concerns and is beneficial just as self-help groups are for others.

Decent housing and jobs that are stimulating and interesting promote a sense of accomplishment and self-worth. Opportunities for recreation and relationships with family and friends are aspects of life that promote mental health as well as physical health as previously discussed. When mental illness is found, evaluation of these areas of a person's life as well as his or her communication skills and opportunity to share concerns may reveal some problem areas. In addition, changes in living arrangements, in staff or care-providers, and in family structure can be stressful and lead to mental health problems. Adults with Down syndrome seem to have a somewhat lower ability to adapt to the stress of such changes. Being aware of the potential for problems and informing the adult with Down syndrome of upcoming changes and allowing time for him to process the information and to express his concerns may lessen the stress and prevent problems.

Physical Health That Can Affect Mental Health

A number of physical health concerns were noted above that can contribute to the psychological or mental decline of an adult with Down syndrome such as hypothyroidism, vision impairment,

vitamin B-12 deficiency and hearing impairment. A thorough evaluation for any of these problems should be performed if mental health is impaired.

Depression

Depression can be difficult to diagnose in persons with Down syndrome and sometimes lasts for years if untreated. It occurs a little more commonly than in the general population. Lack of verbal skills can contribute to the difficulty in diagnosing and treating depression. However, depression is usually treatable and remarkable improvement in daily living skills, motivation and interaction with others is often experienced. Anti-depressants, group or individual therapy and encouragement in participating in daily activities, especially exercise, are beneficial. Sometimes it becomes necessary to involve occupational therapists, respite workers and others to help the patient get moving again.

Obsessive-Compulsive Disorder

Obsessive-compulsive disorder can be seen with depression or on its own and is probably more common in persons with Down syndrome. It will often respond to structuring the environment to reduce the frustration of the compulsion and to medications.

Others

While other psychological problems such as attention deficit hyperactivity disorder and bipolar (manic-depressive) disorder can occur, there is no evidence to suggest that they occur more frequently than in the general population. Schizophrenia is actually thought to be uncommon in persons with Down syndrome.

LABORATORY EVALUATION

In addition to the history and physical, an annual blood test to screen for thyroid disorders is recommended. Hypothyroidism (underactive thyroid gland) occurs in approximately one-third of adults with Down syndrome. The symptoms of hypothyroidism include dry skin, constipation, weight gain, lethargy, depression, cold intolerance, coarsening of the voice, joint aches and muscle cramps. Since a number of these are common characteristics of people with Down syndrome, it can be difficult to diagnose hypothyroidism on the basis of clinical findings. In addition, hyperthyroidism (overactive thyroid) is more common in adults with Down syndrome than in the general population (although much less common than hypothyroidism). Therefore, annual screening for thyroid conditions with a blood test is recommended.

CONCLUSION

Most adults with Down syndrome can live healthy lives. To achieve that goal requires a multi-faceted approach including living a healthy lifestyle, following certain preventive measures, participating in appropriate health screening, and treating health problems early in their course. While there are some disorders that occur more commonly in persons with Down syndrome (and some that occur less frequently), following this approach is a way to promote a healthy life.

From http://www.advocatehealth.com/dl/luth/services/other/adsc/promoting_health.pdf

Healthcare Guidelines for Adolescents and Adults with Down Syndrome – Preventive Medical Check List

1999 REVISION

Edited by William I. Cohen M.D. for the *Down Syndrome Medical Interest Group*¹

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.

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). If a child is below the third percentile, or is 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.

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 augmentative 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.

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References are available online at www.denison.edu/dsq/health99.shtml

Youth and Adult Health Care Guidelines

(Based on 1999 Down Syndrome Health Care Guidelines)*

Adolescence (12-18 Years)

- TSH and T4-Thyroid Function Test (annual).
- Auditory Testing (annually).
- Monitor for obstructive airway; sleep apnea.
- General physical and neurological exam; check for atlanto-axial dislocation. Cervical spine x-ray (as needed for sports).
- Eye examination (annually).
- Monitor for obesity by plotting height for weight on growth charts for typical children
- Clinical evaluation of the heart to rule out mitral/aortic valve problems. ECHO - Echocardiogram (as indicated by findings).
- Reinforce need for subacute bacterial endocarditis prophylaxis (SBE) in susceptible adolescents.
- Adolescent medicine consult for puberty/sexuality issues; health, abuse prevention and sexuality education. Pelvic exam (only if sexually active).
- Low calorie, high fiber diet; regular exercise
- Smoking, drug and alcohol education.
- Begin functional transition planning (16 yrs). Monitor independent functioning.

Adult (18+ years)

- TSH and T4-Thyroid Function Test (annual).
- Auditory testing (every 2 years).
- Cervical spine x-rays (as needed for sports); check for atlanto-axial dislocation. Ophthalmologic exam, looking especially for keratoconus & cataracts (every 2 yrs)
- Clinical evaluation of the heart to rule out mitral/aortic valve problems. Echocardiogram-ECHO (as indicated).
- Reinforce the need for subacute bacterial endocarditis prophylaxis (SBE) in susceptible adults with cardiac disease. Baseline Mammography (40 yrs; follow up every other yr until 50, then annual).
- Pap smear and pelvic exam (every 1-3 yrs. after first intercourse). If not sexually active, single-finger bimanual exam with finger-directed cytology exam. If unable to perform, screen pelvic ultrasound (every 2-3 years). Breast exam (annually).
- General physical/neurological exam. Routine adult care.
- Clinical evaluation for sleep apnea.
- Low calorie, high-fiber diet. Regular exercise. Monitor for obesity.
- Health, abuse-prevention and sexuality education. Smoking, drug & alcohol ed.
- Clinical evaluation of functional abilities (consider accelerated aging); monitor loss of independent living skills.
- Neurological referral for early symptoms of dementia: decline in function, memory loss, ataxia, seizures and incontinence of urine and/or stool.
- Monitor for behavior/emotional/mental health. Psych referral (as needed).
- Continue speech and language therapy (as indicated).

*[HEALTH CARE GUIDELINES FOR INDIVIDUALS WITH DOWN SYNDROME: 1999 REVISION (Down Syndrome Preventive Medical Check List) is published in **Down Syndrome Quarterly** (Volume 4, Number 3, September, 1999, pp. 1-16) and is reprinted, duplicated, and/or transmitted with permission of the Editor. Information concerning publication policy or subscriptions may be obtained by contacting Dr. Samuel J. Thios, Editor, Denison University, Granville, OH 43023 (email: thios@denison.edu).]

Health Care Guidelines - Record Sheet (1999 Revision) Age - 13 Years to Adulthood

Name: _____

Birthday: _____

	Age, in years									
Medical Issues	13	14	15	16	17	18	19	20-29	30-39	40-49
Usual Preventative Care	___	___	___	___	___	___	___	___	___	___
Audiologic Evaluation	___	___	___	___	___	___	___	___	___	___
Ophthalmologic Evaluation	___	___	___	___	___	___	___	___	___	___
Thyroid (TSH & T ₄)	___	___	___	___	___	___	___	___	___	___
Nutrition	___	___	___	___	___	___	___	___	___	___
Dental Exam ¹	___	___	___	___	___	___	___	___	___	___
Sleep Apnea	___	___	___	___	___	___	___	___	___	___
Parent Support	___	___	___	___	___	___	___	___	___	___
Developmental & Educational Services	___	___	___	___	___	___	___	___	___	___
Neck X-rays & Neurological Exam ²	___	___	___	___	___	___	___	___	___	___
Pelvic exam ³				___	___	___	___	___	___	___
Assess Contraceptive Need ³				___	___	___	___	___	___	___

Instructions: Perform indicated exam/screening and record date in blank spaces. The shaded boxes mean no action is to be taken for those ages.

¹Begin Dental Exams at 2 years of age, and continue every 6 month thereafter.

²Cervical spine x-rays: flexion, neutral and extension, between 3-5 years of age. Repeat as needed for Special Olympics participation. Neurological examination at each visit.

³If sexually active.

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Alzheimer's Disease and Down Syndrome

By: Dr. Ira Lott, Professor of Pediatrics University of California at Irvine

Alzheimer's Disease, a degenerative neurological disorder characterized by progressive memory loss, personality deterioration and loss of functional motor capabilities, is far more common in individuals with Down syndrome than the general population. However, not all individuals with Down syndrome will develop Alzheimer's disease, and even those showing Alzheimer's-type symptoms may not have Alzheimer's disease since other conditions can mimic the symptoms.

Q. How common is Alzheimer's disease in individuals with Down syndrome?

A. Estimates vary, but a reasonable conclusion is that 25 percent or more of individuals with Down syndrome over age 35 show clinical signs and symptoms of Alzheimer's-type dementia. The percentage increases with age. In the general population, Alzheimer's disease does not usually develop before age 50, and the highest incidence (in people over age 65) is between five and 10 percent. The incidence of Alzheimer's disease in the Down syndrome population is estimated to be three to five times greater than in the general population.

Q. What are the symptoms of Alzheimer's disease?

A. Early symptoms include loss of memory and logical thinking; personality change; decline in daily living skills; new onset of seizures; changes in coordination and gait; and loss of continence in bladder and bowel habits.

Q. How is a final diagnosis made?

A. Alzheimer's disease is difficult to diagnose. It is important to be certain Alzheimer's-type symptoms do not arise from other conditions, namely thyroid disorders, depressive illness by psychiatric criteria, brain tumor, recurrent brain strokes, metabolic imbalances and various neurological conditions.

The diagnosis of Alzheimer's disease is made on the basis of clinical history, showing a slow, steady decrease in cognitive function and a variety of laboratory tests which provide contributory evidence, including electroencephalogram, brain stem auditory evoked response, computerized transaxial tomography and magnetic resonance imaging, among other tests and measurements.

Q. Is there a baseline test that can be repeated at intervals to determine specific decrease in cognitive function?

A. Psychologists often use questionnaires answered by family members, companions or caregivers that assist in the early detection of dementia. It is recommended that individuals with Down syndrome be tested at age 30 to provide a baseline reading, and periodically thereafter. If the tests show deterioration, further tests must be made to rule out conditions that present similar or overlapping symptoms.

Q. What information has research yielded about a link between Alzheimer's disease and Down syndrome?

A. Current research investigating how certain genes on Chromosome 21 may predispose individuals with Down syndrome to Alzheimer's disease. A number of centers are testing therapies in Down syndrome that appear to benefit patients with Alzheimer's disease in the general population.

Q. How can research into Alzheimer's disease and Down syndrome be advanced?

A. As is true for Alzheimer's disease in the general population, a full understanding of the disorder involves post-mortem examination of brain tissue. Contributions to a brain tissue repository for purposes of extending knowledge about the relationship between Down syndrome and Alzheimer's disease will help to advance research in this area. For information for families and physicians considering such a donation, please contact the National Down Syndrome Society at 800-221-4602.

Summary

- Individuals with Down syndrome are three to five times more likely than the general population to develop Alzheimer's disease. Onset of Alzheimer's may begin as early as age 30 in the Down syndrome population as compared to age 50 in the general population.
- Symptoms of a variety of other diseases and conditions mimic the symptoms of Alzheimer's disease: personality change, decline in daily living skills, memory loss, changes in coordination and gait and other changes. Diseases and conditions such as depression, thyroid disorders, brain tumor, recurrent brain strokes, metabolic imbalances and various neurological conditions must be ruled out prior to a diagnosis of Alzheimer's disease.
- It is recommended that individuals with Down syndrome take a baseline test of cognitive function at age 30, and that this test be repeated annually to determine any deterioration in this function. Some Alzheimer's disease symptoms can be treated, although there is no current means of curing or arresting the disease.
- Current research suggests a causative link between the extra "gene dosage" from the third chromosome 21 of Down syndrome and Alzheimer's disease. To advance research, donations of brain tissue from individuals with Down syndrome and Alzheimer's disease are being sought.

Source: If you would like additional information on Alzheimer's disease and Down syndrome, please refer to the NDSS Resource List on Alzheimer's disease, available online at www.ndss.org or through the NDSS Helpline, (800) 221-4602.

Depression in Persons with Down Syndrome

Can people with Down syndrome have depression?

Just as persons with Down syndrome are as susceptible to illness as is the general population, so are persons with Down syndrome just as susceptible to depression. Specifically, depression in persons with mental retardation has been estimated at 6 to 7%.

What are signs or symptoms of depression?

It is important that any of the signs and/or symptoms of depression be closely assessed to determine if they are symptoms of a biological/medical condition (e.g., seizures) or a condition in itself (i.e., depression). Depression may present differently in persons with Down syndrome. They would be less likely to verbalize feelings of sadness, guilt or worthlessness because of articulation and conceptual limitations. Nevertheless they would show observable changes in mood and behavior. Changes in mood include an increase in irritable mood, lethargy or listlessness. Changes in behavior include withdrawal, loss of interest in activities formerly enjoyed, fatigue and lack of energy, changes in eating or sleeping patterns (either less or more), and there may be a slow down in movement and activities. Self talk may also increase or it may occur in more public settings. These changes may be easily interpreted by caregivers, once they recognize the behavior as depression.

Depression may be missed because the family is told that the signs and/or symptoms are just "the Down syndrome." Signs and/or symptoms might also be misinterpreted as dementia or Alzheimer disease. Parents are primarily the first people to notice these signs/symptoms. It is very important that if any signs/symptoms are present that professional assistance be sought to avoid more serious problems later, such as placement in a psychiatric facility.

What can cause depression in a person with Down syndrome?

Depression can occur as a result of current stress (acute), such as the death of a family member, or develop as a more chronic condition. A recent history of the person's life is necessary in order to define the problem, if possible, and identify available support systems. Assessment must be carried out according to the individual's cognitive/developmental level.

In Down syndrome, as the person ages, mental function and social support may decrease creating the opportunity for depression to occur. Adults with Down syndrome may experience isolation, changes in residences, and the lacking of social opportunities and adaptive skills. All of these factors can lead to decreased self esteem, an inability to care for themselves, and depression. Often it is the case that adults with Down syndrome lack the opportunities to make their own decisions in areas where that are capable (e.g., social activities, hair style, clothing choice, etc.). They may be treated as children and feel little, if any, control over their lives. Individuals who care for adults with Down syndrome may overlook support systems, over protect the person, and/or inappropriately take punitive actions toward the person with Down syndrome.

What can be done to prevent/alleviate depression?

Presently, many physicians, psychologists and other mental health professionals have had little training in psychiatric conditions and mental retardation, leaving room for missed or misdiagnosis. The clinical presentation of depression in persons with Down syndrome may be different from someone without a cognitive deficit. A preventive approach to depression in persons with Down syndrome is suggested. Given the opportunity to take some control over their lives, persons with Down syndrome will experience fewer occasions for depression to occur. It is important that families and mental health professionals be informed about and sensitive to the early signs of depression in persons with mental retardation so that diagnosis and treatment may begin promptly. It is known that more behavioral problems occur when persons are bored and little structure is present in their lives. When early diagnosis is made, it is important that the treatment plan be individualized and follow up available. It is also important that treatment involve the best environment and quality of life possible. When anti depressant medications are necessary, information for the families concerning side effects and the prevention of side effects through periodic blood levels and discontinuation trials are important.

Much more study must go into examining depression in persons with Down syndrome. Understanding why a person feels depressed and alleviating the source(s) is often a challenging endeavor. Successful prevention, intervention and follow up of depression in persons with Down syndrome can best be accomplished through collaboration between the individual, their family and the mental health/medical community.

Prepared for and Approved by the Professional Advisory Council, National Down Syndrome Congress March 1992

Reviewed and Revised February 2006

Menstrual Cycles in Girls with Down Syndrome

By: Marjorie Greenfield, MD, Cleveland, OH

When it comes to menstrual periods, girls with Down syndrome are no different than typical children. The average age for the first menstrual period is around 12 years old. Overweight girls are more likely to menstruate slightly earlier. Most girls begin developing breasts about two years before and have some pubic and armpit hair by the time periods arrive.

For the first few years after menstruation starts, the hormonal system is continuing to mature to adult levels and periods may be quite irregular. Most commonly, early periods occur every three to six weeks and last less than eight days. During this time, girls often don't ovulate (release an egg) and hormonal fluctuations, not ovulation, cause the periods which can be irregular, with changing flow from month to month. The good news is that these sorts of anovulatory cycles don't usually cause bad menstrual cramps or premenstrual syndrome (PMS). Unless the child bleeds excessively, time usually fixes the irregularities.

Over the next five years, cycles usually occur at more regular intervals – 24 to 36 days – and the monthly flow becomes more predictable. Regular monthly periods and cramping are usually signs that the girl or woman is ovulating. At first, the greatest concern for families often is how to prepare their daughters to deal with having periods. It can be frightening to see blood coming from your body without adequate preparation. Girls need to be told what to expect and shown how to use sanitary pads, matched to their level of understanding. After that, families' concerns about menstruation tend to be focused on four areas: menstrual hygiene, menstrual symptoms (cramps, behavior disturbances), sexuality and the possible need for contraception.

Menstrual Hygiene

Although many parents are apprehensive about menstruation, girls who can toilet themselves can be taught to wear and change their sanitary pads. This teaching can be done using the same techniques that were employed for toilet training. If menstruation becomes a big hygiene problem, an occupational therapist with professional experience with these concerns may help. In some situations, medications such as Depo-Provera® or birth control pills can be used to diminish or eliminate menstrual flow.

Menstrual Symptoms

The hormone progesterone, which is released after ovulation (during the two weeks that precede the period), can lead to mood irritability, water retention, constipation and bloating. These symptoms are typically mild, although they may become worse over the years. Placebo-controlled studies have shown that Calcium supplementation, 500 milligrams twice a day for the two weeks before the period, can diminish the physical and emotional symptoms of PMS. Severe premenstrual mood and behavioral disturbances are possible, and can be treated with medications for mood (such as Prozac®), or with hormonal suppression of menstruation. For severe premenstrual behavioral disturbances, seek a gynecological or psychiatric consultation.

Local uterine hormones called prostaglandins cause menstrual cramps. Severe menstrual cramps may cause contractions as severe as those experienced during labor. Unless medical conditions prevent taking these sorts of medications, ibuprofen (Advil® or Motrin®) or naproxen (Aleve®) can help alleviate bothersome menstrual cramps. If cramps are not adequately controlled, a family doctor, adolescent specialist or gynecologist can provide information about

other treatments, including Depo-Provera, Implanon® (a progesterone contraceptive implant that is inserted in the upper arm), the Mirena® intra-uterine device and oral contraceptive pills. My general rule is that no one should miss normal activities due to her periods.

Sexuality

As a parent, it can be difficult to think about your child's sexuality. It may be tempting to think that a child with medical problems or developmental delay doesn't have these reactions and desires. But humans are sexual beings, and girls with DS have sexual feelings just like anyone else. Sexuality issues need to be addressed starting in early childhood, with open discussion about privacy and appropriate touching. When sexual education includes the broadest interpretation of "sexuality" it covers information about all caring relationships, including friendship, love and marriage, addressed at a level the adolescent can understand. Education should cover the differences in male and female anatomy, and types of sexual expression including intercourse. A buddy system, avoiding contact with strangers and only giving out first names can prevent unsafe situations. Puberty is a time when a girl may become vulnerable to sexual advances or sexual abuse. Open conversation, good supervision and learning assertiveness all help to keep girls safe.

Contraception and Sterilization

Even when they do everything to protect their daughters from abuse, some families choose to get their daughters on a birth control method just in case someone takes advantage of her. Many start worrying more about this when their daughter starts menstruating, and the chance of pregnancy becomes real. Many parents have come to me requesting that their daughters get their tubes tied. Laws vary related to sterilization of individuals who cannot consent to their own care, but in most states, laws prevent "forced" sterilization – even when loving parents provide consent. Procedures that permanently prevent pregnancy may not be legally available to a girl or woman who is not capable of understanding the risks, benefits and alternatives and giving legal consent. Typically, a procedure must be necessary for medical reasons and would be offered to a typical woman under the same circumstances, such as a hysterectomy for excessive bleeding.

The Period as a Vital Sign

Menstrual problems are common in young women, and those with DS are no exception. Mild irregularities are normal, especially in the first few years of menstruation. However, these situations may require medical evaluation:

- Periods starting before age nine or ten
- Periods that haven't started by age 16 or three years after breast development
- Going more than three to six months without bleeding
- Bleeding lasting longer than a week
- Cycles that come closer than three weeks apart, counting from the first day of one period to the first day of the next

Menstrual irregularities can be a sign of a medical problem, including hypothyroidism and obesity. Excessive bleeding can also *cause* problems, including anemia (low blood) and social difficulties with managing the flow.

The first step in evaluating abnormal periods is a detailed history. It is very helpful to keep a written record of the periods for a few months, to provide an objective accounting of days of flow. Also, when going to see the doctor, bring a written list of medical conditions and current medications, with dosages. For medicines that commonly interfere with the menstrual

problems, you may want to check with the cardiologist or other medical doctor to find out if there are medical reasons not to use the hormones estrogen or progesterone in this instance. Report any family history of blood clots in the legs (deep venous thrombosis or DVT) or blood clots in the lungs (pulmonary embolism or PE) to the gynecologist.

An internal pelvic examination is not always necessary, and can be omitted if the patient is too fearful or refuses the exam. No one should be forced to have a pelvic examination. For that part of the body, no means no. Over time, with gentle explanations and dividing the exam into manageable parts, most girls accept and tolerate a pelvic exam. But it may not be possible at the first appointment. Other aspects of the physical examination that are helpful in understanding the causes of menstrual problems include weight, hair pattern, abdominal examination, and whether the opening of the vagina looks normal. If the internal organs need to be checked, ultrasound can image the uterus and ovaries. Blood tests are often used to check some hormone levels; and, if bleeding is excessive, to check for anemia. Causes of menstrual problems are categorized into hormonal issues, anatomical conditions and problems with the blood's ability to clot. Medical conditions such as untreated or undertreated hypothyroidism can interfere with female hormones. Likewise, obesity and some medications can cause irregular cycles. Blood thinners may lead to excessive bleeding. In some cases although the flow is technically normal, periods may seem unmanageable. Treating menstrual problems depends on the cause and how the problem affects the patient's life.

Conclusion

Menstruation marks a life transition. For most girls, periods are just something to get used to and manage, just as toileting was earlier in childhood. Your family doctor, pediatrician, adolescent medicine specialist or gynecologist can help sort out normal from abnormal menstruation, and guide you in managing these new challenges.

Source: Connections, Vol. 15 #1, Jan/Feb 2008, newsletter of Down Syndrome Guild of Greater Kansas City

Sexuality in Down Syndrome

By: Dr. William Schwab, MD

Human sexuality encompasses an individual's self-esteem, interpersonal relationships and social experiences relating to dating, marriage and the physical aspects of sex. Sex education, appropriate for the developmental level and intellectual attainment of individuals with Down syndrome, helps in engendering healthy sexuality, preventing unwanted pregnancy and diseases and in alleviating other problems related to sexual function.

Q. Do individuals with Down syndrome have sexual feelings?

A. In the past, sexuality was not considered an issue for young people with Down syndrome because of the inaccurate belief that mental retardation was equivalent to permanent childhood. In fact, all people with Down syndrome do have intimacy needs and sexual feelings, and it is important that these be recognized and planned for in education, residential and other programs and settings.

Q. Do children with Down syndrome develop physically the same way as their peers in the general population?

A. Children with Down syndrome experience the same sequence of physical and hormonal changes associated with puberty as other children their age.

Q. Do children with Down syndrome experience the emotional upheavals characteristic of adolescence?

A. The emotional changes characteristic of adolescence are also present in children with Down syndrome, and may be intensified by social factors. Any adolescent who lives in the community, attends school and is exposed to media inevitably develops an awareness of sexuality. Teenagers and young adults with Down syndrome often express interest in dating, marriage and parenthood. They can be expected to experience typical adolescent changes in mood and outlook.

Q. What kind of sex education is appropriate for individuals with Down syndrome?

A. To be effective, education must be individualized and understandable, focusing not only on the physical reproductive aspects, but also positioning sexuality within the context of all interpersonal relationships. An ideal curriculum will ensure that the individual's understanding of relationships, sexual intercourse and parenting is factual, realistic and socially acceptable.

Q. How can healthy sexuality be encouraged for individuals with Down syndrome?

A. Creating an environment conducive to healthy sexual expression must be considered in designing educational, vocational, recreational and residential programs. Positive sexual awareness can only develop through self-esteem, understanding of social relationships and personal interaction/communication skills. All these factors influence how intimacy needs are met.

Q. Do women with Down syndrome have any special needs or concerns in regard to birth control?

A. Approximately 50 percent of women with Down syndrome are fertile and may use any method of contraception without added medical risk. The method chosen will depend on personal preference, ability to use the contraceptive effectively and possible side effects. Surgical sterilization may also be performed without added risk for women with Down syndrome who are in stable medical condition; however, availability of this procedure to women who are developmentally disabled may be controlled by state law. The individual with Down syndrome should be involved as much as possible in decision-making should this option be considered.

Q. Are there any special needs for individuals with Down syndrome in regard to disease prevention?

A. Men and women with Down syndrome have the same susceptibility to sexually transmitted diseases as the rest of the population. Use of condoms during sexual intercourse is the best known form of protection against AIDS, herpes and other sexually transmitted diseases. Sexual education should include information on sexually transmitted diseases and how to prevent them.

Q. How can a person with Down syndrome be protected against sexual abuse?

A. It is highly recommended that age-appropriate education in protective behaviors begin in childhood and be reinforced throughout the life of the person with Down syndrome. Individuals with Down syndrome must be taught the boundaries of normal physical interactions in the social sphere, as well as the self-assertion skills to enlist help if necessary.

Q. Do girls with Down syndrome have normal menstrual periods?

A. Menstruation for girls with Down syndrome is no different than for their peers in the general population. On the average, they begin menstruating at age 12 1/2, but may begin as early as age 10 or as late as age 14. Most girls with Down syndrome have regular cycles with the same minor irregularities typical of their peer group. Alterations in a previously regular cycle may be due to the normal process of aging, or may be a sign of emerging hyperthyroidism. Ongoing irregularity of menstrual cycle warrants medical examination.

Q. If a woman with Down syndrome becomes pregnant, will the baby have Down syndrome?

A. At least half of all women with Down syndrome do ovulate and are fertile. Between 35 and 50 percent of children born to mothers with Down syndrome will have Trisomy 21 or other developmental disabilities.

Q. When is the onset of menopause for women with Down syndrome?

A. Menopause may occur at a wide range of ages. Typically it takes place after age 40.

Q. Are males with Down syndrome fertile?

A. Scientific information about the fertility of men with Down syndrome is limited. There have

been at least two documented cases where the paternity of a man with Down syndrome was confirmed. It is likely that additional cases will be recognized - especially as more men have the opportunity to live in the community and develop intimate relationships. It is not known if the offspring of men with Down syndrome are more likely to have Down syndrome or other anomalies. It does seem clear that, in general, men with Down syndrome have a significantly lower overall fertility rate than that of other men of comparable ages. An individual's status can be partially assessed by having a semen analysis done, but this may not be definitive. If a couple desires pregnancy prevention, contraception should always be used.

Q. Do boys with Down syndrome mature later than their peer group in the general population?

A. The onset of puberty in boys may be slightly delayed, but this is not a major factor. Genital anatomy is comparable to that of boys who do not have Down syndrome.

Summary

- People with Down syndrome experience the same range of sexual feelings as the general population.
- Teenagers with Down syndrome undergo the same changes at puberty as all adolescents, though these changes may be slightly delayed for boys with Down syndrome.
- While the overall fertility of men with Down syndrome may be significantly reduced, it is still advisable that couples use contraception whenever pregnancy prevention is desired.
- At least 50 percent of women with Down syndrome are fertile. Healthy females can use contraception without added medical risk.
- Education is an appropriate and highly desirable component in developing positive sexual awareness for the individuals with Down syndrome.

If you would like additional information on sexuality and Down syndrome, please refer to the NDSS Resource List on sexuality, available online at www.ndss.org or through the NDSS Helpline, (800) 221-4602.

About the Author: Associate Professor Department of Family Medicine and Practice University of Wisconsin

Source: ©1995-2003 National Down Syndrome Society, (800) 221-4602, www.ndss.org

Sleep Apnea

By: Paula Cho

In order to understand what sleep apnea is, it is important to know what normal sleep is. Normal, uninterrupted sleep consists of a cyclic pattern alternating between rapid eye movement (REM) sleep and non-rapid eye movement (non-REM) sleep. REM sleep is also called dream sleep. During REM sleep there are many physiological changes observed. For instance, there is a decline in chin muscle activity, generalized muscle atony (relaxation) except the diaphragm, and irregular breathing. Consequently, with the onset of normal sleep, the pharynx (airway) narrows because of muscle relaxation, causing added resistance to air movement through the airway. During normal sleep, this causes reduced air movement and a slight increase in carbon dioxide in our body.

What is sleep apnea? Sleep apnea is defined as a complete cessation of breathing from any cause during sleep, resulting in decreased oxygen in the blood or increased carbon dioxide (a greater increase than would be seen in normal sleep). The pauses in breathing usually last 10 to 20 seconds but can last as long as 120 seconds. In severe cases, more than 500 attacks of sleep apnea may occur during a night.

Sleep apnea is divided into 2 major types. The most common is obstructive sleep apnea, which is caused by obstruction of the airway. This is the type that is believed to be more common in people with Down syndrome. Respiratory effort continues but the obstruction prevents movement of air into and out of the lungs. The less common type is non-obstructive (central) sleep apnea. In this type, there is a problem in the brain or nervous system that controls our breathing. The airway may be open and the muscles may be fine but the signal to breathe is not sent or doesn't reach the muscles.

Sleep apnea is increasingly recognized as a significant cause of health problems and even death. More than one percent of the general population is affected, with a dramatic increase in the elderly. Approximately 4% of middle-aged men have obstructive sleep apnea whereas women are much less frequently affected. Obesity is the major risk factor although it is not invariable (many obese people do not have sleep apnea and some people with sleep apnea are not obese). Typically, conditions that narrow the upper airway, such as enlargement of the soft palate, uvula, tonsils, adenoids, local fat deposition, enlarged tongue and other structural abnormalities predispose to sleep apnea.

Additionally, persons with Down syndrome are predisposed to obstructive sleep apnea because their relatively small midfacial region with a relatively large tongue contributes to the compromised airway. Narrowing of the opening of the skull for the nasal passages and narrowing in the airway below the pharynx may also contribute to a diminished airway. Chronic inflammation of the nasal passages, enlarged lymph tissue (including the tonsils and adenoids) and obesity, which are common in persons with Down syndrome, are added risk factors. Maintenance of an open airway is dependent on the muscle tone in the pharynx. Muscle tone in this area is often reduced in persons with Down syndrome as part of a generalized decline in muscle tone. The use of sedative medications and, perhaps, antihistamines may also contribute to decreased muscle tone.

Characteristically, a long history of loud snoring combined with restless sleep, excessive daytime drowsiness, and early morning headaches suggest the diagnosis of sleep apnea. Also

there may be an inability to concentrate, depression, irritability and personality changes. During obstruction, there may be aspiration of secretions into the lungs causing a cough or aggravating asthma symptoms. As the problem progresses, shortness of breath and fatigue increase.

The diagnosis of apnea begins with recognition of the presenting symptoms. Hypothyroidism (underactive thyroid) should be excluded. The physical exam usually, but not always, reveals obesity and excessive soft tissue in the mouth, pharynx and neck. With advanced disease, the right side of the heart is weakened and findings of failure of that part of the heart may be seen. The laboratory tests are usually normal except low oxygen and high carbon dioxide may be found.

An overnight sleep study is needed to definitely diagnose sleep apnea. It involves recording eye movements, muscle tone, electroencephalogram (EEG) to measure brain waves, and electrocardiogram (EKG) to measure the electrical activity of the heart. The test also records respiratory movements, nasal and oral air movement, and oxygenation of the blood.

If obstructive sleep apnea is found, there are several alternatives for treatment. If the patient is overweight, the first consideration is to lose weight. Many people have less or no sleep apnea if they don't sleep on their back. Therefore, encouraging the person to sleep on their side or on their abdomen can be helpful. Sometimes placing a sock on the back of the pajama top and putting a tennis ball in the sock keeps a person off their back. Eliminating sedatives and alcohol also helps some people.

Oxygen by a nasal cannula (the two-pronged device commonly used in the hospital to deliver oxygen) can help some individuals. However, generally if the oxygen is to get to the lungs, oxygen alone is not enough. For those individuals, CPAP (continuous positive airway pressure) is used. This is delivered by a mask that fits over the nose or the mouth and nose. It delivers a positive pressure to the airway to keep it open.

For some individuals, surgery to open the airway is indicated. Removal of nasal polyps, correction of a deviated septum or removal of enlarged tonsils and adenoids can sometimes help. However, often there is also obstruction in other parts of the upper airway and a more extensive surgery is required. Uvulopalatopharyngoplasty is a surgery that removes the uvula, redundant soft palate tissue, tonsils, and adenoids. When there is no other available treatment, some patients require a tracheotomy (hole in the neck) to allow breathing around the obstructed airway.

If sleep apnea is not treated, serious complications can arise. In addition, the chronic sleep deprivation of sleep apnea and the poor oxygenation can lead to significant mood and behavioral changes that can be misinterpreted as purely psychological. Care must be taken in these instances because a medication with sedative effects may be prescribed to help with these changes. The sedation could make the sleep apnea worse which could cause an increase in mood and behavior disturbance.

People with Down syndrome have a higher incidence of sleep apnea. Awareness of the possibility of sleep apnea and observation for the symptoms of daytime drowsiness, disturbed sleep, and pauses in breathing while sleeping are the first steps towards making the diagnosis.

<http://www.advocatehealth.com/luth/services/other/adsc/publications.html>

Sleep Hygiene

David Duff, MD

Family Practice Resident at Lutheran General Hospital

As many of you are already aware, women and men with Down syndrome have a much higher rate of Obstructive Sleep Apnea, a disorder causing decreased quality of rest due to halted breathing while asleep. It can lead to excessive daytime sleepiness, chronic fatigue, and possibly more serious health problems.

While it is important to be aware of this potential sleep problem, something that is often overlooked (by people with and without Down syndrome) is the day-to-day sleeping habits known as “sleep hygiene”. Just as good dental hygiene involves a set of planned activities such as brushing and flossing your teeth, choices and plans you make during the day can affect how healthy your night’s sleep ends up being. It is estimated that about half of all patients who come to doctors’ offices admit, when asked, that they have some type of problem sleeping.

One night without good sleep does not have much affect on our performance the following day. Add together a string of bad nights and you start seeing effects. It is hard or impossible to stay awake during the day (especially for “boring/repetitive” tasks), tempers are shorter, creativity suffers, and you can start feeling “run down” all the time. Here are some suggestions for ways to improve the quality of normal sleep - to have good sleep hygiene:

1. Set a daily bedtime and a wake-up time. Stick to this schedule, even on weekends.
2. Exercise daily, but not within an hour of bedtime if possible. Late afternoon may be the best.
3. No caffeine after lunchtime. It would be best to avoid it altogether (it stays in your system 12 to 15 hours), but this habit is hard to break. Caffeine includes coffee, black tea, soda pop, and, yes, chocolate. Herbal tea is O.K.
4. Minimize sleep interruptions-keep noise/light levels at an absolute minimum (snoring roommates can be a problem); make sure the room is not too hot or cold; don’t drink a lot of water before bedtime; have a comfortable bed.
5. No big meals within 1-2 hours of bedtime. A light snack is O.K.
6. No smoking---ever.
7. Wind down toward the end of the day. No challenging or upsetting activities (if possible) in the evening. This can mean setting aside time earlier in the day for thinking through difficult issues or problems.
8. Avoid taking naps during the day, especially in the evening. If you do nap, nothing longer than 20 to 30 minutes.
9. Use your bed only for sleeping. No TV, reading, homework, eating, etc.

As with all people, there will be differences between individuals on the amount of sleep needed (6 to 9 hours per night seems to be the average), what activities help or hinder sleep, and just how much chocolate actually counts as a caffeine risk. It is also normal to have sleeping problems when going through difficult or stressful times in life. If the disturbances continue for weeks, however, it is time to get help.

The direct impact of Down syndrome on sleep is not fully known. There are some characteristics that many with DS may find keep them from sleeping as they could. A group home environment or having roommates may lead to more disruptions of sleep. Sorting through emotional,

complex, and stressful issues may take more time and effort leading to increased time lying awake “just thinking”. Regular exercise is also lacking for some adults with Down syndrome.

Finally, as every human ages, there are expected changes in sleep habits. Staying asleep is more difficult; older adults wake up more frequently during the night and early morning. The amount of deep sleep is less than in young adults. With these changes comes an increase in daytime sleepiness and napping. Other medical problems can begin to interrupt sleep and the natural body sleep cycles become irregular.

If you suspect someone you know needs help with their sleep hygiene, start keeping track of bedtimes, wake-up times, and the other issues listed above. Try to make some of the sleep hygiene modifications to fit your particular needs.

Good night!

<http://www.advocatehealth.com/luth/services/other/adsc/publications.html>

Speech and Language Resource Guide for Adolescents with Down Syndrome

By Libby Kumin, Ph.D., CCC-SLP, *Founder*, The Center for the Study of Speech and Language in Children with Down Syndrome

Why can't my adolescent communicate?

The most common communication problems for adolescents with Down syndrome are in the areas of:

- speech intelligibility, i.e. that their speech may be difficult to understand
- conversational skills, i.e. they have difficulty with long conversations
- narrative discourse, i.e. difficulty telling about what happened to them or retelling a story
- academic language, i.e. abstract language for subject learning in middle school and high school and
- asking for specific clarifications when they don't understand something.

What type of speech and language evaluation and treatment are needed?

What can I do to improve my child's ability to communicate?

In middle school and high school, communication skills need to support where the child is now (subject learning, following school rules and routines, and social language skills) and also support planning for the future (language for transition planning, job skills and community living). Classroom language skills become more difficult because your child has multiple teachers who have a variety of teaching and language styles. In middle and high school, academic subjects involve more abstract and advanced language and there are fewer contextual cues and visual models to help learning. During adolescence, if some communication skills cannot be mastered, alternate communication solutions may need to be considered. For example, if the adolescent cannot give his vital information so that it can be understood (name, address, telephone number, date of birth), business cards or an electronic communication device can be used to ensure that he is able to give that information when needed. All of the following areas should be assessed, and treated as needed:

Receptive Language Skills

- Advanced Comprehension
- Vocabulary and concept development
- School, transition to workplace
- Auditory memory
- Auditory processing
- WH questions
- Following complex directions (written and verbal)
- Literacy (reading skills)

Expressive Language Skills

- Vocabulary
- Morphology
- Syntax
- Answering questions
- Giving vital information
- Encoding/sentence formulation
- Presentation skills

Pragmatics/Social Language Skills

- Requests
- Social interactive skills
- Conversational skills
- Narrative discourse
- Clarifications and repairs
- School to job language skills

School Language Skills

- Language of the Curriculum
- Language of Instruction
- Language of the Hidden Curriculum
- Language of Testing
- Language of Classroom Routines
- School to Job Transition Skills

Speech Skills

- Articulation
- Phonology
- Oral motor skills
- Childhood apraxia of speech (if needed)
- Speech intelligibility
- Voice
- Fluency
- Rate of Speech

Where can I go for help?

Books, conferences and presentations will help you learn more about how to help your adolescent with speech and language. Speech and language services may be available through your local school system through the IEP process and transition planning, medical centers, community clinics, university clinics, and private practitioners. Your local parent support group is an excellent source of information to help in locating professionals in your area.

- Kumin, L. (2006). *What Did You Say: A Guide to Speech Intelligibility in Children with Down Syndrome*. Bethesda, MD: Woodbine House.
- Kumin, L. (2003). You Said It Just Yesterday, Why Not Now? Developmental Apraxia of Speech in Children and Adults with Down Syndrome. *Disability Solutions*, 5 (2), 1-15. (on-line www.disabilitysolutions.org)
- Kumin, L. (2002). Maximizing Speech and Language in Children and Adolescents with Down Syndrome, in Cohen, W., Nadel, L. & Madnick, M. (Eds). *Down Syndrome: Visions for the 21st century* (403-415).NY:Wiley-Liss.
- Kumin, L. (2002). Why Can't You Understand What I am Saying: Speech Intelligibility in Daily Life. *Disability Solutions*, 5, 1-15. (www.disabilitysolutions.org)
- Kumin, L. (2001). *Classroom Language Skills for Children with Down Syndrome: A Guide for Parents and Teachers*. Bethesda, MD: Woodbine House.

For more information:

- www.ndscenter.org
- www.ndss.org
- www.ds-health.com
- www.downsed.org

This work made possible by a grant from The Thomas W. Bradley Foundation.
<http://www.ndscenter.org/resources/documents/speech/Kumin.Adolescents.pdf>

Speech and Language Resource Guide for Adults with Down Syndrome And Their Parents

By Libby Kumin, Ph.D., CCC-SLP, *Founder*, The Center for the Study of Speech and Language in Children with Down Syndrome

Why can't my adult with Down syndrome communicate?

Learning to communicate is an ongoing process. The vast majority of adults with Down syndrome use speech to communicate. The most common communication problems for adults with Down syndrome are that their speech may be difficult to understand (speech intelligibility) and that they have difficulty with long conversations, with telling about what happened to them or retelling a story, and with asking for specific clarifications when they don't understand something.

What type of speech and language evaluation and treatment are needed?

For adults with Down syndrome, communication skills need to support their daily lives at home, at work, and in the community. Although very few adults go for speech and language therapy, research has shown that adults can improve their speech and language skills. One innovative treatment method used watching soap operas in a group therapy setting as a means to improve listening skills, conversational speech, and retelling stories.

Adults with Down syndrome who are working in the community need to be able to navigate the transportation system and ask for help if necessary, talk and socialize with co-workers, understand and follow directions at work. Their speech needs to be understandable to other people. It is helpful if they are able to use communication technology such as voice mail and email.

In expressive language, areas that can be evaluated and worked on in therapy or at home are vocabulary, sentence formulation, answering questions, and giving vital information. The adult needs to be able to make requests and to ask for and offer clarifications when the message is not understood. Adults with Down syndrome usually do well with social interactive language. They use greetings, and scripts (automatic phrases such as "Hi!" and "See you later!") effectively. They may have difficulty with conversational skills, and tend to have short conversations or rambling conversations that veer from the topic. In the area of speech, articulation, phonology, oral motor skills, childhood apraxia of speech can be evaluated and treated if needed. Other factors that affect speech intelligibility such as stuttering and voice problems sometimes occur in adults. Self talk and vocal mannerisms such as clearing the throat also occur in adults. It is felt that self talk is used to review and process what has occurred during the day.

What can I do to improve the communication skills for an adult with Down syndrome?

Specific communication difficulties can be addressed in speech therapy at any age. For adults, practice with communication skills is important. Adults who are advocates and have opportunities for public speaking often improve their communication skills. Social activities, job-related communication, volunteer or other community group activities, trips, celebrations and

other events provide opportunities for people with Down syndrome to practice their communication skills.

Where can I go for help?

The National Down Syndrome Congress has an annual convention that includes information and social activities for adults with Down syndrome. Books and websites are other sources for information.

- Chicoine, B. & McGuire, D.(2006). *Mental Wellness in Adults with Down Syndrome*.
- Bethesda, MD: Woodbine House.
- Pueschel, S. (2006). *Adults with Down syndrome*. Baltimore, MD: Brookes Publishing.
- www.ndsccenter.org
- www.ndss.org
- www.ds-health.com
- www.downsed.org

This work made possible by a grant from The Thomas W. Bradley Foundation.
<http://www.ndsccenter.org/resources/documents/speech/Kumin.Adults.pdf>

"Self-Talk" in Adults with Down Syndrome

By: Dennis McGuire, Ph.D., Brian A. Chicoine, MD. and Elaine Greenbaum, Ph.D..

Do you talk to yourself? We all do at different times and in various situations. In examining and evaluating over 500 patients at the Adult Down Syndrome Center of Lutheran General Hospital, we have heard repeatedly that adults with Down syndrome talk to themselves. Sometimes, the reports from parents and caregivers reflect deep concern that this behavior is "not normal" and symptomatic of severe psychological problems.

Preventing misinterpretation of self-talk as a sign of psychosis in adults with Down syndrome is a major motivation for this article. Too often, we believe, these conversations with self or imaginary companions have been equated with "hearing of voices" and treated with anti-psychotic medications (such as Haldol, Mellaril, or Risperdal). Since it is extremely difficult to evaluate the thought processes of adults with cognitive impairments and limited verbal skills, we urge a very cautious approach in interpreting and treating what seems to be a common and at times very helpful coping behavior for adults with Down syndrome.

The Adult Down Syndrome Center (the Center) was developed to address the health and psycho-social needs of adults with Down syndrome. The Center is a unique collaboration of the National Association for Down Syndrome, a parent advocacy group, Advocate Health Care, a private health care provider, and the Institute on Disability and Human Development at the University of Illinois at Chicago. Since its inception in 1992, over 500 individuals have been evaluated at the Center.

Our records at the Center indicate that 81 percent of adults seen engage in conversations with themselves or imaginary companions. Patients have ranged in age from 11 to 83 years of age. The median age in our data base is 34. This high prevalence of self-talk does not seem to be widely known. For some parents and caregivers, the fact is reassuring. But the content of these conversations, their frequency, tone, and context can be important in determining if treatment is warranted.

Helpful Self-Talk

Families and caregivers should understand that self-talk is not only "normal" but also useful. Self-talk plays an essential role in the cognitive development of children. Self-talk helps children coordinate their actions and thoughts, and seems to be an important tool for learning new skills and higher level thinking. Three-year-old Suzy says to herself: "This red piece goes in the round hole." Then Suzy puts the red piece into the round hole of the puzzle.

We suspect that self-talk serves the same useful purpose of directing behavior for adults with Down syndrome. Consider the case of twenty-two-year-old "Sam" (not his real name). His mother reported the following scene. She asks Sam to attend a family function on a Sunday afternoon. Sam's regular routine is to go to the movies on Sunday afternoons. Sam tells his mother he will not go with the family. Then the mother asks Sam to think it over. Sam storms off to his room and slams the door. His mother overhears this dialogue:

*"You should go with your family, Sam."
"Listen to your Mom!"
"You can go next Sunday."*

*"But I want to go to the movies."
"But Sunday is my movie day."*

Sam's mother said he went to the family function, with the proviso that he could go to the movies the next Sunday. Sam may have been talking to an imaginary person or arguing with himself, but Sam clearly managed to cope with a situation not to his liking.

In children without identified learning problems, the use of self-talk is progressively internalized with age. Moreover, children with higher intellectual abilities seem to internalize their self-talk earlier. As self-talk is transformed into higher level thinking, it becomes abbreviated and the child begins to *think rather than say* the directions for his or her behavior. Thus, the intellectual and speech difficulties of adults with Down syndrome may contribute to the high prevalence of audible self-talk reported to us at the Center.

In general, the functions of self-talk among adults are not as well researched or understood. Common experience suggests that adults continue to talk to themselves out loud when they are alone and confronting new or difficult tasks. Though the occurrence may be much less frequent, the uses of the adult's self-talk seem consistent with the findings about children. Adults talk to themselves to direct their behavior and learn new skills. Because adults are more sensitive to social context and may not want to overhear these private conversations, their self-talk is observed less frequently.

Adults with Down syndrome show some sensitivity about the private nature of their self-talk. Like Sam in the example above, parents and caregivers report that self-talk often occurs behind closed doors or in settings where the adults think they are alone. Having trouble judging what is supposed to be private and what is considered "socially appropriate" also may contribute to the high prevalence of easily observable self-talk among the patients visiting the Center.

In the general population, self-talk among older persons is frequently notable and, usually, easily accepted, just as it is with children. Among the elderly, social isolation and the increasing difficulty of most tasks of daily living may be important explanations for this greater frequency of self-talk. For adults with Down syndrome, these explanations also make sense. Adults with Down syndrome are at greater risk for social isolation and the challenges of daily living can be daunting.

Additionally, we have found that many adults with Down syndrome rely on self-talk to vent feelings such as sadness or frustration. They *think out loud* in order to process daily life events. This is because their speech or cognitive impairments inhibit communication. In fact, caregivers frequently note that the amount and intensity of the self-talk reflects the number and emotional intensity of the daily life events experienced by the individuals with Down syndrome.

For children, the elderly, and adults with Down syndrome, self-talk may be the only entertainment available when they are alone for long periods of time. For example, a mother reported that her daughter "Mary" spent hours in her room talking to her "fantasy friends" after they moved to a new neighborhood. Once Mary became more involved in social and work activities in her new neighborhood, she did not have the time or the need to talk to her imaginary friends as often.

Thus, that adults with Down syndrome use self-talk to cope, to vent, and to entertain themselves should not be viewed as a medical problem or mental illness. Indeed, self-talk may be one of the few tools available to adults with Down syndrome for asserting control over their lives and improving their sense of well-being.

When to Worry

The distinction between helpful and worrisome self-talk is not easy to cast in stone. In some cases, even very loud and threatening self-talk can be harmless. This use for self-talk by the adult with Down syndrome may not be that different from someone who rarely swears but screams out a four-letter word when hitting her thumb with a hammer. Such outbursts may simply be an immediate, almost reflexive outlet for some of life's frustrations.

Our best advice about when to worry is to listen carefully for changes in the frequency and context of the self-talk. When self-talk becomes dominated by remarks of self-disparagement and self-devaluation, intervention may be warranted. For example, it may be quite harmless when "Jenny" yells "I am a dummy," once, right after her failure to bake a cake from scratch. However, if Jenny begins to tell herself over and over "I am a dummy and can't do anything right," it may be time to worry and to do something. A marked increase in the frequency and a change in tone of the self-talk also may signal a developing problem. For example, a caregiver reported that "Bob" had begun to talk to himself more frequently and not just in his room at the group home. Bob seemed to lose interest in his housemates and spent more time in these conversations with himself. Bob talked to himself, sometimes loudly and in a threatening manner, at the bus stop, at the workshop and at the group home. Bob was diagnosed as experiencing a severe form of depression. Over an extended period of time, Bob began to respond to an anti-depressant and to his participation in a counseling group.

In another case, "Jim" (like Bob) showed a dramatic increase in self-talk. Jim refused to go to his workshop and to participate in the social activities that he once enjoyed. It turned out that Jim's change in behavior was not due to depression. Instead, Jim's family and staff at his workshop discovered that Jim was being intimidated and harassed by a new co-worker. With the removal of the bully from his workshop, Jim gradually regained his sense of trust in the safety of the workshop. His self-talk and interest in participating in activities returned to earlier levels.

Further study of the content, context, tone, and frequency of the self-talk of adults with Down syndrome may provide more insight into their private inner worlds. What we have observed and heard from family and caregivers suggests that self-talk is an important coping tool and only rarely should it be considered a symptom of severe mental illness or psychosis. A dramatic change in self-talk may indicate a mental health or situational problem. Despite the odd or disturbing nature of the self-talk, our experience at the Center indicates that self-talk allows adults with Down syndrome to problem-solve, to vent their feelings, to entertain themselves, and to process the events of their daily lives.

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The Groove

By: Dennis McGuire, PhD., Director of Psychosocial Services at the
Adult Down Syndrome Center, Lutheran General Hospital

One of the most interesting and consistent findings from the Adult Down Syndrome Center is the discovery that people with DS need sameness, repetition and order in their lives. We call this tendency the “groove” because thoughts and actions of people tend to follow fairly well worn paths, or grooves.

Types of Grooves

One of the most common grooves is a set pattern or routine in one's day to day activities.

Many will often have set methodical and meticulous morning routines for dressing and grooming, daily work routines, as well as evening routines for relaxing and preparing for bed. We have also found that many individuals are meticulous in the care of their rooms and possessions. Much time and effort is spent in making things “just so”. Beds are often made and rooms are neat and tidy. Closets and drawers are quite often in perfect order. In addition, people often have a set place for furniture and other personal items in their rooms or living spaces. Parents and caregivers have found that if any of these items are moved or disturbed by others they will be returned to the original location in short order. A wide array of other grooves also exists. The most common centers are personal preferences for such things as music, sports teams or celebrities. Grooves may also include such personal issues as a favorite relative or a love interest. Also common are grooves which have independence issues as a theme. These are often expressed as “I want to do it (a particular activity) by myself and in my own way”. Some people may also develop self-absorbing grooves around emotionally charged experiences. These may involve positive experiences, such as an award, or negative experiences, such as the loss of a loved one.

Advantages of Grooves

There are numerous advantages to grooves. They give an important sense of order and structure to peoples' lives. They also help persons, who process things more slowly in a fast moving world, have some control over their lives. Routines help to organize and manage daily living tasks which increase independence. Once an activity is learned and becomes part of a daily routine, there is rarely a need for “prompting” or supervision from others.

The ability to follow routines at set times each day can be of great benefit. For example, independence is greatly enhanced when a person is able to get up and off to work on his own. Independence and performance may also be enhanced in the work environment. Employers are often impressed with an employee's reliability in completing routine work tasks and in adhering closely to work time schedules.

Grooves may also offer a refuge from the stresses and strains of daily life. This may be especially important to persons with DS because communicating with others may be tiring and frustrating due to expressive language limitations. In dealing with daily stresses, people will often repeat a specific, enjoyable activity in a quiet or private space.

In the worksite, many people will also schedule time during their daily routine, or when needed, to be alone. The most common and often only place for privacy in the work site is the washroom.

Grooves involving the conscientious care of one's appearance, room and personal items can be of great benefit. Such care of one's appearance conveys an image of pride, self respect and dignity, which will often help develop a good self image. Meticulous care of ones room and possessions may also increase pride and self respect. As an added benefit, family members and caregivers in other settings who share living spaces often appreciate this kind of groove.

Finally, and most importantly, the groove is a powerful means of expression and communication. This is especially true for people with DS, who have limited ability to express themselves verbally. Each groove is a clear and unambiguous statement of a personal choice or preference. For example, daily grooves and routines express how people choose to organize and manage such things as the care of their own grooming, appearance and personal items, their participation in social, recreational, and work activities, as well as personal preferences in music, hobbies and artistic endeavors. Each person's choices will in turn help to shape and define their own unique style and personality.

Disadvantages and Minor Problems

Although there are many benefits and advantages, there are also some disadvantages to grooves that sometimes cause problems. Some of the problems need not be serious if handled appropriately by caregivers. For example a person may be interested in a particular issue, such as a favorite sports team, which they retell over and over to family and friends. While this may be a minor irritant to caregivers, it is not necessarily a problem that interferes with important spheres of living. Additionally there are grooves that may be adaptive if done at the appropriate time or place. For example, a groove for cleaning the bathroom may be greatly appreciated by family members unless it is done in the morning when everyone in the family needs to prepare for work. Similarly, a restaurant manager may be pleased with clean washrooms unless patrons have to wait for long periods while a meticulous job is done.

At the Adult Down Syndrome Center we have also found that a person's need for order or sameness may ironically clash with their need for meticulousness and cleanliness. This is because some people prefer to wear the same shirt or comfortable pair of jeans, over and over, rather than a stiff new pair. Similarly, others may fold and put away dirty clothes rather than have them sit in a rumpled pile in the laundry basket.

More Serious Problems

On the other hand, a groove may become a maladaptive rut when it interferes with functioning in the important spheres of living. There are a number of ways in which this may happen. Some persons may become inflexible about the completion of grooves and routines that may interfere with their participation in other important life activities. For example, one may rigidly adhere to an evening room cleaning and organizing groove rather than choose an opportunity for social or recreational activities which may be beneficial to their health and well being. Some may also make poor decisions, which then become "bad habits". For instance, we have seen a number of people who have serious problems with sleep deprivation. This may happen if one gets in the habit of staying up late to watch movies or TV, even though he or she must get up early for work or school. Others may acquire unhealthy food habits, such as the intake of too much junk food or soda. Some may get stuck on a particular issue such as a love interest, a favorite celebrity, or

the loss of a close relative. Others may get stuck on certain behaviors, which are either part of a regular routine, such as housekeeping tasks, or activities outside one's normal routine, such as flushing toilets or turning lights on and off.

Additionally, the benefits of a groove, derived from sameness, order, and repetition, may become a serious disadvantage in the face of change. For example, gains to one's independence from following a set routine at a set time may be lost if the person is not able to adjust to inevitable changes that occur in daily life. For instance, some may have difficulty changing a morning routine to prepare for an earlier arrival of the bus to school or work. In the workplace people may have trouble adapting to changes in the schedule or in doing activities which are not part of their regular routine.

Apart from daily changes, grooves may also create serious problems for people when they are confronted with major life changes or events, such as transition from school, a move into a residential facility, or the loss of a parent or primary caregiver. These changes often interrupt the bedrock of established grooves and routines which people use to manage their daily lives. In response to these changes some will persist in following old routines or they may get stuck on a particular issue or behavior.

Serious problems may also result if caregivers misinterpret a person's need to complete routines or grooves as oppositional behavior. For example, many persons with Down syndrome try to finish a routine before starting a newly assigned task. Unfortunately, if the care provider believes the motivation for delaying the new task is to resist authority, then an escalating conflict may ensue. Pressure by the care provider may cause further entrenchment by the individual with Down syndrome.

A similar problem may occur if the rules in a residential facility interfere with the completion of grooves. For example, in an attempt to be fair, many group home policies specify that residents can only do a particular housekeeping task for one week. As you might expect, some would much prefer to do the same task for extended periods rather than to change every week. Predictably, conflicts occur when the staff tries to enforce this rule. We have encountered these types of problems most often in residential settings with care providers who have had little experience with persons with DS. We have also found similar problems in workshop settings. We believe this misinterpretation of the groove is one of the reasons people with DS have a reputation for being "stubborn".

Recommendations to Caregivers

When faced with a person seemingly stuck in a groove, we recommend that caregivers first identify and minimize any possible stressors or precipitants to the problem. In some cases, reducing stress may be sufficient to free up someone that is stuck. In other cases action by caregivers may still be necessary after stress is reduced.

When action is needed, caregivers should be careful with how they approach the person who is apparently stuck. Attempts to directly force the person having difficulty with a groove will usually worsen the problem. Caregivers who are most successful at helping in these circumstances, understand the need and benefits of grooves. Instead of a direct and forceful approach, they will slowly and gently help the person resolve the issue in a positive way. This approach will encourage new steps which will become the basis of a new, more productive groove.

Caregivers also need to be mindful that problems with grooves may masquerade as behavior problems. This may occur if a groove is misinterpreted as oppositional behavior.

We have found that once caregivers understand and accept the fact that grooves can be helpful, problems are more easily resolved. The tension and hostility derived from misinterpretation of a groove and an ensuing power struggle will often be replaced by a more conciliatory and cooperative interaction between caregiver and the person with DS. Parents and other care providers who seek professional help for a person dealing with a stuck groove need to choose a professional who has experience in working with persons with DS. Professionals may be experts about certain things but parents have a lifetime of experience and are experts on their son or daughter. In our experience parents' instincts are usually accurate and they understand the grooves that their family member has developed. This is also true for other caregivers that have had a long relationship with the individual. Be sure that the professional listens carefully and looks at all areas of the person's life (health, significant changes in social, school or work environments or other stressors) for possible causes of the problem. Be sure that the professional does his or her homework before making a diagnosis.

Finally, there are a number of preventive measures which may reduce the chance of problems. One of the best ways to learn to deal flexibly with changes is to have persons with DS begin early in life to gain mastery over small day to day changes and challenges.

The earlier and more consistently these opportunities are presented, the more likely people will be able to adapt to life changes when they occur. In this way, individuals develop a comfortable pattern or groove for dealing with change. Another important preventive measure is for parents and other caregivers to attend school or workplace staffings to ensure that others have a clear understanding of the nature and benefits of grooves.

From www.nads.org

Weight Management in Down Syndrome: The Adult Years

By: Joan E. Medlen, R.D.

Weight management is something that everyone would benefit from understanding early in life. The first article in this three-part series looked at the importance of the feeding relationship to a healthy lifestyle and early eating habits for young children with Down syndrome. The second article looked at concerns of the school-age and adolescent years: carefully building independence and modeling the habits that shape a healthy attitude toward food and activity. This article, the last of the series, continues to focus on issues that are relevant to weight management and Down syndrome, with an emphasis on concerns that arise in adulthood. For adults with Down syndrome, weight management involves more than the physical changes of maturity. It is a complex mix of community involvement, friendships, daily routines, living situations, activity, and food choices along with the physical challenges of age. This article will explore many of these influences and how we, as parents, family, and support people can encourage adults with Down syndrome toward a healthy lifestyle.

The Effects of Metabolism

Research suggests that children with Down syndrome have a lower basal metabolic rate (BMR) than their same-aged peers. BMR is the rate a person burns calories for fuel when completely at rest—or sleeping. As a result, a child with Down syndrome uses fewer calories while they are asleep and also throughout the day. In addition, adults with Down syndrome (ages 18-20) have finished growing and require fewer calories than they did as a child. If eating or exercise habits do not change to compensate for this decrease in energy requirements, the energy equation quickly becomes unequal with Calories in outweighing Calories out.

Slowing or Stopping the Rate of Weight Gain

If your child is still gaining weight consistently, focus on stopping or slowing that upward trend. This is a quick fix, however, and only meant as a detour while the two of you consider other factors and make changes. To do this, it is important that your adult child understands and shares your concern regarding his weight gain and overall health. Without him on your team, you will be waging a losing battle. Working together, success is far more likely.

The first thing to do is to visit with your family doctor to rule out any medical causes of weight gain such as hypothyroidism. Be certain to ask if there are any physical limitations to be considered if your child decides to increase his physical activity. Once the physician has given you the "green light" for lifestyle changes, your child and you can begin to create a plan together for a healthy lifestyle.

To slow or stop weight gain, begin by choosing an area of the energy equation to modify. Keep in mind you're not trying to accomplish weight loss at this time. Focus on beginning healthful habits that your adult child is willing to do. Some areas to consider are:

- Increasing activities such as walking, and using stairs,
- Confining eating to designated areas,
- Balancing meals,
- Planning snacks rather than eating uncontrolled, and
- Menu planning.

Usually small changes in any one of these areas will slow or stop consistent weight gain. Do not try to create perfect habits. Accept and encourage any changes, no matter how small, your child is willing to make. Many times the most effective initial change to encourage is an increase in activity. A walk around the block for each 30 minutes of television watched is very effective in our culture. Increasing activity has some other health benefits that may spill over to other areas as well. They include, but are not limited to:

- An increase in positive attitude,
- An increase in metabolic rate after exercise,
- A decrease in appetite,
- An increase in lung capacity,
- A decrease in resting heart rate,
- An increase in muscle mass and muscle strength, and
- A decrease in blood pressure.

Once you have both agreed on a plan to slow or stop weight gain, it is time to move on to the next step: evaluating and shaping opportunities for choice and control regarding food and activity.

Making Lifestyle Changes: Who Decides?

In the past, when a medical intervention was needed for a person with Down syndrome, a professional stepped in to create a plan or a treatment to correct the situation. Basically, weight management was done to them. This can happen in any living situation, and still does. Many well-meaning parents and professionals deny adults with Down syndrome control over their food choices by mandating a strict menu, a dietary restriction, or an exercise regime. This rarely leads to long-term success in weight management. In fact, if the person with Down syndrome is not involved in the decision-making process, it might lead to rebellious choices such as covert eating or uncontrolled eating at social gatherings in an effort to regain control of food choices. For this reason, it is wise to avoid "diets" and "treatment plans" by involving the person with Down syndrome in discussions about weight and overall health promotion and letting them take the lead in the decision-making process.

One way to begin identifying potential areas for change is to evaluate the living situation of your adult child and try to create opportunities for choices and control that include healthy options as much as possible.

In Group Settings:

Meet with the owner or manager of the living group to discuss your concerns. Consider asking the following questions:

- Are the people who live in the group home involved in menu planning? Cooking? Shopping?
- Does the menu structure allow for more than one entrée and a variety of side dishes for each meal?
- Does the support staff model and encourage healthy choices?
- Is there a variety of healthy foods available for snack choices?
- Are opportunities for physical activity such as low-impact aerobics, walking, or biking available? How often?

Once you have asked these questions, share the options with your child. Emphasize that he is in control of his options by pointing out the areas he has choices to make.

Living at Home with Family:

Ask yourself this question: Is better health a priority for everyone who lives in your home? It is important to send a consistent message regarding healthy habits to your child with Down syndrome. If it is, gather as a family to discuss what changes each person would like to make for a healthy lifestyle. Most families do not have weekly menus, but now would be a good time to begin using them.

Together, create menus for meals and snack choices. Most meals (breakfast and lunch in particular) can be written to include different options for varying needs in the family. The point of the menu is to ensure that only foods the family agrees upon are in the home. Involve your adult child in cooking and shopping activities.

Supported and Independent Living:

Menus are very helpful for adults with Down syndrome living independently. However, they must be written by the self-advocate. Encourage using a variety of foods in menu planning. When cooking for one or two, or when cooking is a lot of work, it is easy to only make a few things that are favorites like macaroni and cheese, peanut butter sandwiches, and other easy-to-prepare dishes. One way to encourage experimentation with new recipes is to plan a dinner together once a month. The theme of the meal is trying new recipes. It is easier to try something new with a companion. Together, you can choose a new recipe to try, create the menu for the night, shop for what is needed for the recipe, provide support to your child learning the recipe, and live the adventure of tasting this meal together. If it is good, then begin adding it to the menu once a month.

In this living situation, parents and support people have the least influence or control over what actually happens. However, menu planning, when done by the person with Down syndrome with support, only as needed, not only provides structure, but also makes shopping and budgeting much easier because it is planned in advance. By planning ahead using a menu, the foods needed for recipes are on the shopping list, and therefore available when it's time to cook them. Menu planning is the best defense against developing the habit of standing in front of the cupboards trying to decide what to cook. Cupboard-side menu planning typically leads to fast foods or skipped meals.

Nutrition Guidelines: A "Rule of Thumb" Approach

As a parent or support person, it is helpful to have an idea of the most basic concepts for healthy eating. Use these guidelines to assist you:

- Plan for and eat three meals a day.
- Time those meals so they are not more than 5 hours apart. If meals need to be longer than 5 hours apart due to work or school schedules, plan for a snack.
- Each meal should consist of 3 out of the 5 food groups for overall balance.
- Each planned snack should consist of 2 out of the 5 food groups for overall balance.
- Don't go out of your way to plan for the "sometimes/others" group (the sixth group at the top of the pyramid). They seem to find their way into menus on their own.

- Encourage your child to take one-half of the portion they feel like eating. When that is gone, have them set the timer for 15 minutes. If they are still hungry after the timer goes off, then seconds are in order.

These guidelines will ensure overall nutritional balance throughout the day while leaving room for food preferences and individual decisions. Teaching adults with (or without) Down syndrome to choose within these parameters because they are healthy for everyone will promote healthful choices without feeling restrictive. If you are worried about the nutrients that may be missed by food choices, an over-the-counter vitamin will take care of any nutrients that are missed.

Activity & Friendships: Essential Pieces of the Puzzle

The last area to consider, though just as important, is activity. What kind of opportunities does the adult with Down syndrome have to create new friendships, socialize with old friends, and participate in recreational activities? In a recent study, researchers found a correlation between friendships and opportunities for recreation and the body mass index (BMI) of adults with Down syndrome. BMI is a measurement used to assess body weight and health risks using body weight and body fat. This study suggests that for adults with Down syndrome, friendships and social interactions have a more consistent and direct effect on BMI than diet and exercise. This is a very strong message. It suggests that better health and weight management for adults with Down syndrome must include the element of friendships and social recreation in addition to the usual healthy lifestyle choices (eating and exercise) to be successful. Friendships and opportunities for recreation are "inextricably linked" to overall health.

As a dietitian and mother of a child with Down syndrome, I see this as an opportunity to create some healthful options for people with and without Down syndrome now and in the future. Once again, it will require advocacy by parents, parent groups, and professionals to be successful. Some places to consider beginning include:

- Joining walking or Volksmarch clubs, especially those associated with a hospital,
- Offering community aerobic classes at the group home or nursing home,
- Doing aerobic tapes with friends at home or in apartments,
- Joining local health clubs and buying YMCA memberships,
- Hiring a personal trainer at a health club or YMCA to work with two or three adults with Down syndrome as a small group,
- Joining a hiking group for beginners,
- Creating an exercise buddy system with friends and relatives, or
- Walking the dog with a group of dog owners.

Although it is important to promote healthy eating habits, the nutrition and activity pieces to the puzzle of weight management for adults with Down syndrome are relatively easy. The harder pieces to fit into place are the ones we cannot dictate or force: choosing to eat wisely, choosing to be active, and cultivating the friendships that complete the puzzle of a fulfilling and healthy life for our children. The best we can do is to ensure the tools for a healthy lifestyle are taught, available, and encouraged in a positive way, and to encourage friendships and recreation throughout their lives.

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