



Are there organizations or websites that will help me when I have more questions?

Can you tell me what some of those abbreviations mean?

Organization Acronyms

AAA - Area Agency on Aging – *advocates on behalf of member agencies for services and resources for older adults and persons with disabilities. By providing a range of options that allow older adults and persons with disabilities to choose the home and community-based services and living arrangements that suit them best, AAAs make it possible for older adults to remain in their homes and communities as long as possible.* - www.n4a.org (to search for your local office)

AAIDD – American Association on Intellectual and Developmental Disabilities - *promotes progressive policies, sound research, effective practices, and universal human rights for people with intellectual and developmental disabilities.* – www.aidd.org

AIDD - Administration on Intellectual and Developmental Disabilities - *dedicated to ensuring that individuals with developmental disabilities and their families are able to fully participate in and contribute to all aspects of community life in the United States and its territories.* - www.acl.gov/programs/AIDD

APSE - Association of People Supporting EmploymentFirst - *advances employment and self-sufficiency for all people with disabilities through advocacy and education.* - www.apse.org

The Arc - (formerly Association for Retarded Citizens) - *promotes and protects the human rights of people with intellectual and developmental disabilities and actively supports their full inclusion and participation in the community throughout their lifetimes. Their website offers resources for health, financial planning, residential options and more.* - www.thearc.org

AUCD - Association of University Centers on Disabilities - *a network of interdisciplinary centers advancing policy and practice for and with individuals with developmental and other disabilities, their families, and communities.* - www.aucd.org

DD Council - Developmental Disabilities Council (each state has one) - *develops and sustains inclusive communities and self-directed services and support for individuals with developmental disabilities.* - www.nacdd.org (to search for your state office)

DSAIA – Down Syndrome Affiliates in Action - *supports the growth and service capabilities of local and regional Down syndrome organizations.* – www.dsaia.org

EEOC - Equal Employment Opportunity Commission - *responsible for enforcing federal laws that make it illegal to discriminate against a job applicant or an employee because of the person's race, color, religion, sex (including pregnancy), national origin, age (40 or older), disability or genetic information. It is also illegal to discriminate against a person because the person complained about discrimination, filed a charge of discrimination, or participated in an employment discrimination investigation or lawsuit.* - www.eeoc.gov

HHS - Health and Human Services – *the principal agency for protecting the health of all Americans and providing essential human services, especially for those who are least able to help themselves. Provides millions of children, families, and seniors with access to high-quality health care, by helping people find jobs and parents find affordable child care, by keeping the food on Americans' shelves safe and infectious diseases at bay.* - www.hhs.gov/iea/regional/index.html (to search for your regional or state office) or www.aspe.hhs.gov/office_specific/daltcp.cfm (for the office of disability, aging and long-term care policy)

Organization Acronyms

IMDSA – International Mosaic Down Syndrome Association - *supports any family or individual whose life has been affected by Mosaic Down syndrome by continuously pursuing research opportunities and increasing awareness in the medical, educational and public communities throughout the world* - www.imdsa.org

NARIC - National Rehabilitation Information Center - *conducts comprehensive and coordinated programs of research and related activities to maximize the full inclusion, social integration, employment and independent living of individuals of all ages with disabilities.* - www.naric.com

NDRN – National Disability Rights Network - *the nonprofit membership organization for the federally mandated Protection and Advocacy (P&A) Systems and Client Assistance Programs (CAP). Collectively, the P&A/CAP network is the largest provider of legally based advocacy services to people with disabilities in the United States.* – www.ndrn.org

NDSC - National Down Syndrome Congress - *provides information, advocacy and support concerning all aspects of life for individuals with Down syndrome.* – www.ndscenter.org

NDSS - National Down Syndrome Society - *advocates for the value, acceptance and inclusion of people with Down syndrome.* - www.ndss.org

NIH – National Institute of Health - *the largest source of funding for medical research in the world. NIH is made up of 27 centers and institutes, including the NICHD which conducts and supports laboratory research, clinical trials, and epidemiological studies that explore health processes; examines the impact of disabilities, diseases, and variations on the lives of individuals; and sponsors training programs for scientists, health care providers, and researchers.* - www.nih.gov

ODEP – Office of Disability Employment Policy - *develops and influences policies and practices that increase the number and quality of employment opportunities for people with disabilities.* - www.dol.gov/odep

SLN – Sibling Leadership Network - *provides siblings of individuals with disabilities the information, support, and tools to advocate with their brothers and sisters and to promote the issues important to them and their entire families.* – www.siblingleadership.org

SSA - Social Security Administration – *pays disability benefits under two programs, SSI and SSDI* – www.ssa.gov

TASH - *an international advocacy association of people with disabilities, their family members, other advocates, and people who work in the disability field.* – www.tash.org

The State of the States in Developmental Disabilities Project - *administered by the University of Colorado and funded in part by the AIDD, it provides longitudinal data for 1977-2011. The new State Profiles each present six pages of data on I/DD spending, revenue and other program trends in each state and DC. Customized, state-by-state comparative data, also recently updated through 2011, are also available.* - <http://www.stateofthestates.org/>

Common Disability Acronyms

ABLE act - Achieving a Better Life Experience
(H.R. 647)

AD - Alzheimer's Disease

ADA - Americans with Disabilities Act

ASD – Autism Spectrum Disorder

AT - Assistive Technology

BD - Behavior Disorder

CBI - Community Based Instruction

CM - Case Manager

DD - Developmental Disabilities

DME - Durable Medical Equipment

DS - Down Syndrome

FERPA - Family Education Rights and Privacy
Act of 1974

HHA - Home Health Agency

HSA - Home Service Agency

I/DD – Intellectual/Developmental Disabilities

ICF - Intermediate Care Facility

IDEA - Individuals with Disabilities Education
Act

IFSP - Individualized Family Service Plan

IHP - Individual Habilitation Plan

IL - Independent Living

ILC - Independent Living Center

ILP - Integrated Living Plan

IRWE - Impairment-Related Work Expenses

ISP - Individual Service Plan

LOC - Level of Care

OT - Occupational Therapy/Therapist

PT - Physical Therapy/Therapist

PWD/PWDs - People with Disabilities

RT - Recreation Therapist

Sib – Sibling

SILP - Semi-Independent Living Program

SLP - Speech Language Pathologist

SLT - Speech Language Therapist

SMI - Supplementary Medical Insurance

SNT – Special Needs Trust

SSDI - Social Security Disability Insurance

SSI - Supplemental Security Income

TANF - Temporary Assistance to Needy Families

VR - Vocational Rehabilitation

Bibliography

FOR PARENTS & ADULT SIBLINGS

McGuire, Dennis and Brian Chicoine. *Mental Wellness in Adults with Down Syndrome*. Bethesda, MD: Woodbine House. 2006.

Directors of the Adult Down Syndrome Center in Illinois share their experience in working with more than 3000 patients since 1992. This book addresses specific disorders and diagnoses, and offers treatment ideas for both professionals and caregivers.

McGuire, Dennis and Brian Chicoine. *The Guide to Good Health for Teens & Adults with Down Syndrome*. Bethesda, MD: Woodbine House. 2010.

In their second book the doctors address specific health care concerns as individuals with DS age. This includes an in depth look at Alzheimer's disease and DS.

Pueschel, Siegfried M. (Ed.). *Adults with Down Syndrome*. Baltimore: Brookes Publishing. 2006.

A comprehensive book on the social, clinical, legal, and personal issues faced by adults with Down syndrome. The book blends contributions by professionals who work with adults with DS along with personal essays by self-advocates. Topics include: medical and mental health, employment, post-secondary education, social relationships and living arrangements.

TRANSITION PLANNING

Bigby, Christine. *Moving on Without Parents*, Baltimore: Brookes Publishing. 2000.

This book provides resource and support for planning transitions out of the family home and preparing for the phase of life in which parents are not the caregivers for the middle-aged and older adults with Developmental disabilities.

Simons, Jo Ann. *The Down Syndrome Transition Handbook: Charting Your Child's Course to Adulthood.*, Bethesda, MD: Woodbine House. 2010.

Simons writes from a professional perspective as someone who worked with many families as they went through Transition. As a mother, she has lived it. The book explains legal issues, funding mechanisms, options for "creating a meaningful and purposeful day" once the school bus stops coming, housing, post-secondary education opportunities, health care and more.

BOOKS FOR SIBLINGS

Dougan, Terrell Harris. *That Went Well: Adventures in Caring for My Sister*. New York, NY. Hyperion, 2008.

Between her sister Irene's spirited behavior and Terrell's firm commitment to giving Irene the least restrictive life she can lead, Terrell's great sense of humor is a tremendous gift. Caring for Irene was a family commitment — and a big one at that due to explosive temper outbursts — and provided lots of useful fodder for entertaining stories that share life lessons.

Bibliography

Gallagher, Peggy, Thomas Powell and Cheryl Rhodes. *Brothers & Sisters: A Special Part of Exceptional Families*. 3rd edition. Baltimore: Brookes Publishing. 2006.

The subject of this book is about the relationship of siblings, where one has a disability, as they move into adulthood.

McHugh, Mary. *Special Siblings: Growing Up With Someone With a Disability*. Baltimore: Brookes Publishing. 2002.

Reflections on her own life growing up with a brother with cerebral palsy and mental retardation, the author also interviews more than one hundred other siblings of individuals with special needs.

Meyer, Donald and Emily Holl. *The Sibling Survival Guide: Indispensable Information for Brothers and Sisters of Adults with Disabilities*. Bethesda, MD: Woodbine House. 2014.

A guidebook for managing the care and support of a sibling with a disability.

Meyer, Donald. *Views from Our Shoes: Growing Up With a Brother or Sister with Special Needs*. Bethesda, MD: Woodbine House. 1997.

A collection of essays by children and young adults who have a sibling with special needs.

Meyer, Donald and Patricia Vadasy. *Living with a Brother or Sister with Special Needs: A Book for Sibs*. Seattle: University of Washington Press. 1996.

Discusses specific disabilities in easy to understand terms, and the intense emotions brothers and sisters experience.

Meyer, Donald. (Ed.) *Thicker Than Water: Essays by Adult Siblings of People with Disabilities*. Bethesda, MD: Woodbine House. 2009.

Thirty-nine adult siblings wrote essays about their experiences growing up with their now adult siblings with disabilities. They write of relationships that are complex and simple, joyful and hard, accepting and resentful, ordinary and extraordinary.

Simon, Rachel. *Riding the Bus With My Sister*. Boston: Houghton Mifflin, 2002.

A memoir, the author commits herself to spending one day a week for a whole year with her sister, who has developmental disabilities. It was an eye-opening experience in which Simon realizes how little she knew about her sister's life.

HEALTH ISSUES FOR ADULTS WITH DOWN SYNDROME

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Health is more than the absence of disease. Health is a sense of physical, mental and spiritual well being. It is a process that involves health promotion, health monitoring, and early intervention for health problems. Understanding what is typical or in the normal range for a person with Down syndrome is essential for providing health care.

I. Understanding Normal/Typical

Adults with Down syndrome have a number of typical behavioral issues that are important to understand so as not to over-diagnose disease states.

- A. Self-talk and imaginary friends
These appear to be developmental stage-appropriate behaviors that are used as coping strategies, defense mechanisms, and to alleviate boredom.
- B. The Groove
People with Down syndrome often show a tendency towards needing sameness, repetition and order in their lives. It can be very functional.
- C. Grief
A delayed response to grief is often seen and it may be demonstrated in alternative ways.
- D. Pain Tolerance
People with Down syndrome may have an increased tolerance to pain. However, limited communication skills may also limit the ability to

express/convey pain that can lead to the pain being expressed in alternative ways.

- E. Behavioral change as a potential communication device
Sometimes a behavioral change may be an attempt to communicate physical or psychological discomfort. A thorough medical evaluation is indicated when a person with DS presents with a behavioral change to assess for an underlying physical condition.

II. Health Promotion

- A. Regular exercise. We have found that adults with Down syndrome are more likely to be closer to their ideal body weight if they have opportunities for recreational and social activities (not necessarily exercise). We generally recommend 20-30 minutes of exercise at least 3-5 times per week. In addition, (or alternatively) social activities like shopping, visiting museums, etc. that involve walking can be very beneficial. Increasing energy expenditure by parking a little farther away, taking the stairs, and working in the house and yard are all beneficial. Generally, turning off the television and being more active is beneficial.
- B. Recreational Activities - As noted above, recreational activities can have a benefit for physical health. In addition, they are an important part of mental health as well as part of life's enjoyment. Sometimes as parents naturally slow down with age or the adult with Down syndrome ages out of the school system, fewer activities are available. That is a particularly important time to seek other reliable people who can assist in participation in recreational activities.
- C. Nutrition – Obesity is the most common nutrition-related disorder. Attention to a healthy diet as well as regular activity and exercise are required to prevent and treat obesity.
- D. Opportunities for Accomplishment and Sense of Worth – Adults with Down syndrome have the same need as others to feel a sense of accomplishment and worth. For some, that may be a repetitive job that fulfills their need for order and regimen. For others, it may be a particular sense of being needed achieved through doing for others. An assessment of what the individual would like to get from a job, as well what his skills are to do the job is encouraged.
- E. Immunizations – (These recommendations assume all childhood immunizations were given appropriately.)
 - Diphtheria – Tetanus- Pertussis– recommended if no dT in 10 years, then resume dT every 10 years
 - Influenza – consider annually each fall especially if exposure to many people. Recommended annually for persons with certain other health problems.
 - Pneumonia – We recommend considering the pneumonia vaccine at age 50 for adults with Down syndrome. Recommended at a

younger age for persons with certain other health problems and then repeat at age 50.

Varicella (Chicken Pox) – recommended testing for immunity by blood tests if there is no history of having had chicken pox. Recommend the 2-shot series if not immune.

Hepatitis B – recommended for people living in group facilities. We also recommend it for persons working in a group setting (e.g. workshop) and considering it for all others. We recommend a blood test before administering the vaccine for adults with Down syndrome to see if they have immunity (thus, they do not need the vaccine). In addition, we recommend drawing a blood test (hepatitis B surface antibody) to document attaining immune status 4 to 6 weeks after the third shot.

F. Osteoporosis prevention-Osteoporosis is more common in adults with Down syndrome. Adequate calcium intake throughout life is essential. Recommend 1000 mg a day of calcium for men and non-menopausal females and 1500 mg a day for menopausal females (by diet or supplement). Recommend taking Calcium with vitamin D to promote absorption. Consider bone density scanning to screen for osteoporosis. Consider appropriate medical therapy for prevention or treatment of osteoporosis. Consider checking Vitamin D level (blood test).

III. Health monitoring

A. Health Screening – History and Physical exam recommended annually.

B. Some important aspects of the history:

- Decline in skills
- Memory impairment
- Swallowing difficulties, choking
- Change in gait, unsteadiness
- Incontinence of urine and/or stool
- Change in appetite
- Change in weight
- Behavioral issues
- Psychological concerns
 - Change in mood
 - Change in interest in life

C. Thyroid – Recommended annual thyroid blood testing.

D. Celiac Disease – Consider blood testing (anti tissue-transglutaminase antibody IgA).

E. Neck x-ray – consider every 10 years (lateral cervical spine xray in flexion, extension and neutral) to assess for cervical subluxation/atlando-axial instability.

F. Cancer of the cervix – Recommend pap smear every 3 years (after two annual tests normal) if not sexually active and asymptomatic.

G. Breast cancer – Recommend discussing with your medical practitioner. Consider a mammogram annually after 40. Annual breast exam and teach/encourage breast self-exam (if possible).

H. Cancer of the testicle – Recommend annual testicular exam and teach self-exam (if possible).

I. Vision – Recommend exam every 1-2 years.

J. Hearing – Recommend audiogram every 1-2 years.

K. Sleep apnea- sleep apnea is common in people with DS. Annually review history of snoring, pauses in breathing, and restless sleeping. However, many people will have sleep apnea even when not observed by others. Sleep apnea can also cause mood changes, behavioral changes, and a decline or change in skills. Order sleep study if indicated by history.



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Health conditions associated with aging and end of life of adults with Down syndrome

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Abstract

Expectations for the life course of individuals with Down syndrome (DS) have changed, with life expectancy estimates increasing from 12 in 1949 to nearly 60 years of age today (Bittles & Glasson, 2004; Penrose, 1949). Along with this longer life expectancy comes a larger population of adults with DS who display premature age-related changes in their health. There is thus a need to provide specialized health care to this aging population of adults with DS who are at high risk for some conditions and at lower risk for others. This review focuses on the rates and contributing factors to medical conditions that are common in adults with DS or that show changes with age. The review of medical conditions includes the increased risk for skin and hair changes, early onset menopause, visual and hearing impairments, adult onset seizure disorder, thyroid dysfunction, diabetes, obesity, sleep apnea and musculoskeletal problems. The different pattern of conditions associated with the mortality of adults with DS is also reviewed.

Keywords

Down syndrome; physical health; aging; mortality

A highly significant change in the survival of people with Down syndrome (DS) has occurred during the last two generations. In the 1940s, the average life expectancy for individuals with DS was 12 years (Penrose, 1949). With medical breakthroughs and improvements in services, individuals with DS now enjoy life expectancies into their 60s (Bittles & Glasson, 2004). As a result, we are now witness to the first generation of individuals with DS who have benefited from a revolution during their lifetime of better knowledge, health care, advocacy and services (Yang, Rasmussen & Friedman, 2002). Along with this longer life expectancy comes a larger population of adults with DS who display premature age-related changes in their health. There is thus a need to provide specialized health care to this aging population of adults with DS who are at high risk for some conditions and at lower risk for others. Adults with DS are at age-related increased risk for dementia, skin and hair changes, early onset menopause, visual and hearing impairments, adult onset seizure disorder, thyroid dysfunction, diabetes, obesity, sleep apnea and musculoskeletal problems. Along with these increased risks for some conditions comes a different pattern of conditions associated with the mortality of adults with DS. This review focuses on the rates and contributing factors to medical conditions that are common in adults with DS or that show changes with age.

Skin and hair changes

Adults with DS experience a number of dermatological and autoimmune symptoms characteristic of accelerated aging, including premature graying of the hair, hair loss and wrinkling of the skin (Lott, 1982). Significant differences in the skin and chronological age of the individual have been found in post-mortem examinations of the skin of adults with DS

(Edwards, 1978). Further, sun-exposure may have a greater effect on skin wrinkling for individuals with DS than for the general population, contributing to the appearance of premature skin aging in adults with DS (Brugge, Grove, Clopton, Grove & Piacquadio, 1993).

Alopecia areata (the loss of hair) is estimated to effect between 6% to 18.4% of adults with DS (duVivier & Munro, 1975; Prasher, 1994b). In comparison, only 0.1% of individuals with intellectual disability (ID) and 0.1- 0.2% of the general population are affected by alopecia areata (Safavi, Muller, Suman, Moshell & Melton, 1995; duVivier & Munro, 1975). It is posited that this elevated rate of alopecia areata in adults with DS may be related to immunological deficiency in thymus dependent function (Carter & Jegasothy, 1976).

Other skin conditions common in adults with DS include atopic dermatitis, fungal infections, seborrhoeic dermatitis, and xerosis, affecting 34% to 39% of adults (Kerins, Petrovic, Bruder & Gruman, 2008; Prasher, 1994b; Roizen & Patterson, 2003). These skin conditions are found in higher rates among older than younger adults with DS, and may be due in part to declines in self-care associated with memory loss and dementia that are common in the aging of adults with DS (Kerins et al., 2008).

Several theories have been proposed to account for the premature aging observed in adults with DS. The DNA damage theory of aging focuses on aging as a consequence of unrepaired DNA damage accumulation. The DNA damage theory is supported by lower levels of DNA repair enzymes being found in adults with DS (Madan, Williams & Lear, 2006). An alternate theory focuses on free-radical metabolism which involves a key enzyme, CuZn superoxide dismutase, which is located on chromosome 21 (Druzhyna, Nair, LeDoux & Wilson, 1998). Over-expression of this enzyme leads to the altered structure and function of tissue (Sinha, 2005).

Menopause

Women with DS are found to experience menopause starting at an earlier age than other women with ID and than women in the general population (Carr & Hollins, 1995; Cosgrave, Tyrrell, McCarron, Gill & Lawlor, 1999; Schupf, Zigman, Kapell, Lee, Kline & Levin, 1997). An early report found that 87% of women with DS and 69% of women with ID had stopped menstruating by age 46. All women with DS had stopped menstruating by the age of 51, and all of the women with ID by age 54. An estimate of the median age at menopause for women with DS of 47.1 is two years younger than the estimated median age for women with ID of 49.3 (Schupf et al., 1997). These estimates among women with DS and ID are potentially underestimates as the proportion of women who had never menstruated in the sample was unknown. A recent prospective study using multiple methods of analysis of the age at menopause for women with DS, all of whom had a history of menstruation, reported a median age of 45.8 to 47.1 (Seltzer, Schupf & Wu, 2001). No woman with DS older than 52 was menstruating. In comparison, the age at menopause in the general population is 51.3, with perimenopause starting at 47.5 (McKinlay, Brambilla & Posner, 1992), indicating that women with DS have a median age of menopause that is 4 to 6 years earlier than women in the general population.

The earlier onset of menopause in women with DS has implications for their health as menopause is a risk factor for heart disease, depression, osteoporosis, breast-cancer and dementia in the general population (Harlow & Ephross, 1995), and is associated with cognitive declines and dementia in women with DS (Patel, Seltzer, Wu & Schupf, 2001; Schupf et al., 2003). Some studies have found thyroid deficiency contributing to earlier onset of menopause (Carr & Hollins, 1995), whereas others have not replicated the findings in part due to appropriate thyroid treatment being available to the women sampled (Schupf

et al., 1997; Seltzer et al., 2001). As such, the contribution of thyroid conditions to earlier menopause is still uncertain.

Vision impairments

Visual impairments (44-71%) and eye abnormalities are common among aging adults with DS (Gardiner, 1967; Jacobson, 1988; McCarron, Gill, McCallion & Begley, 2005). The prevalence of visual impairments does not appear to be related to level of intellectual functioning among individuals with DS, and is greater than the prevalence (8-50%) observed in older adults with ID without DS (Gardiner, 1967; Haveman, Maaskant & Sturmans, 1989; Janicki & Jacobson, 1986; Moss, 1991). Comparable to visual acuity in the general population, visual impairment deteriorates among adults with DS, with the prevalence of severe visual impairment increasing from 18% among 30-39 year olds, to 28% among 40-49 year olds and to 45% among 50-59 year olds (Van Buggenhout et al., 1999). Of greater concern than the prevalence of visual impairment are reports of only 53% of cases of impairment being diagnosed, and only 50% of individuals with DS receiving correction for their visual impairment (Jacobson, 1988).

Cataracts (11-33%), strabismus (23-37%), refraction problems (30-34%) and keratoconus (15%) are common ophthalmological problems (Aitchison, Easty & Jancar, 1990; Prasher, 1994b; Van Allen, Fung & Jurenka, 1999; Van Buggenhout et al., 1999). A small study of 19 adults with DS over the age of 40 reported even higher rates of ophthalmological problems, with 55% suffering from strabismus and 75% from refraction problems (Hesmes, Sand & Fostad, 1991). The prevalence of cataracts is greater in adults than in children with DS and is comparable to or higher than the prevalence of 17% found in adults in the general population (Congdon et al., 2004). Higher rates of cataracts among adults with DS have been reported (30-68%), with few warranting surgery (Hesmes et al., 1991; Pueschel, 1990; Van Allen et al., 1999). Even when surgery is performed, visual problems and impairments may persist related to aphakia (absence of the lens of the eye) and to poor use of bifocals (Van Allen et al., 1999). Senile cataracts appear to develop earlier in adults with DS than in the general population and to increase in prevalence with age (Pueschel, 1990; Robb & Marchevsky, 1978; Van Allen et al., 1999). These senile cataracts, characterized by a gradual thickening of the lens, tends to occur at a younger age in DS compared to other individuals with ID, possibly due to the accelerated aging process present in individuals with DS and the increased amounts of free radical reactions (Ellis, 2002).

Keratoconus also increases in prevalence with age, from 11% in middle-age to 20% in the elderly (Van Allen et al., 1999). One small study reported that 37% of adults over the age of 40 had keratoconus (Hesmes et al., 1991).

Hearing Impairments

Age-related hearing loss is more common among adults with DS compared to the general population, and appears to have an earlier age of onset. High frequency sensorineural hearing impairments (such as presbycusis) in adults with DS onsets about 20 to 30 years earlier than in their peers with ID, and about 30 to 40 years earlier than in the general population (Buchanan, 1990). Rates of hearing loss among adults with DS range from 12 to 72% (Howells, 1989; Prasher, 1994b; Van Buggenhout et al., 1999), and may depend on the nature of the hearing assessment. Using detailed audiometric methods, Van Buggenhout and colleagues (1999) found 53% of adults with DS to have moderate hearing loss, 17% to have a severe hearing loss, and 2% to have a profound hearing loss. These rates of hearing loss were reported to increase with age and for the hearing loss to become more severe (Buchanan, 1990; Van Buggenhout et al., 1999). While the rate of hearing loss is not related

to level of ID, it has been found to be higher among adults with co-morbid dementia (McCarron et al., 2005).

Howells (1989) reported that 55% of adults over age 21 may have sensorineural hearing loss. For adults over 35 years of age, 68% of ears tested were reported to have cochlear hearing loss, with conductive hearing loss found in 5% of ears and mixed hearing loss found in 13% of ears (Evenhuis, Van Zanten, Procaar & Roerdinkholder, 1992). Van Buggenhout and colleagues (1999) similarly found a low rate of conductive hearing loss (10%) in their sample of adults with DS, and a comparable number with sensorineural and mixed hearing loss (45% and 44% respectively). However, Evenhuis et al. (1992) argues that we do not know how much the rate and degree of hearing impairment in adults with DS is related to previous middle ear pathology, such as otitis media. With the life expectancy of adults with DS growing longer, it is important to understand how medical conditions common early in the life course may influence the health of the individual later in life.

While hearing impairments are common among adults with DS, few are identified or treated. Of individuals identified with hearing impairments during research testing who did not use hearing devices, 80% of their care providers and general practitioners were unaware of the hearing loss (Van Buggenhout et al., 1999). Additionally, only 41% of individuals with hearing impairments identified during research testing were using hearing devices (Van Buggenhout et al., 1999). Half of this sample had severe to profound ID which may have contributed to a difficulty in using hearing devices.

Seizure disorder

The rate of seizures increases with age for individuals with DS, especially for individuals suffering from comorbid dementia. Early reports of the rate of seizures were significantly lower than more recent prevalence estimates. One early estimate of the rate of seizures was 12.2% for adults with DS over the age of 55 and 15.8% for adults over the age of 60 (Veall, 1974). However, early estimates were consistent with more recent estimates that abnormal readings reflective of seizure activity are high, with 71.4% of adults over 55 showing this abnormal activity (Tangye, 1979). Although these early reports underestimated current prevalence estimates, perhaps consistent with the shorter life expectancy of the time, these studies consistency reported a rise in the rate of seizures with age from adolescents to young adults to later adulthood. This increase in the rate of seizures across the lifespan is supported by other later research. Cohort differences show that 8% of adolescents or young adults with DS suffer from seizures as compared to 24-28% of seniors aged 50 and older (Johannsen, Christensen, Goldstein, Nielsen & Mai, 1996; McDermott, Moran, Platt, Wood, Isaac & Dasari, 2005). McVicker, Shanks and McClelland (1994) also supported a lower rate of seizures among younger individuals (7%) as compared to adults older than 50 (46%). A lifespan study of individuals with DS found that approximately 8% of individuals with DS suffer from seizures, and of these 40% experienced seizures after the age of 20 (Pueschel, Louis & McKnight, 1991). The higher rate of seizures observed in individuals with DS may be related to the gene for myoclonus epilepsy being mapped to chromosome 21, however this form of seizure is more commonly found in children and adolescents (Hattori et al., 2006). Older patients with DS typically have tonic-clonic (formerly known as grand mal), complex partial or simple partial seizures (Pueschel et al., 1991). Alternatively, structural abnormalities and biochemical aberrations of the CNS in adults with DS may in part be responsible for increased seizure frequency (Pueschel et al., 1991).

The increase in seizures related to age parallels the increase in the general population, although the rate of seizures is lower in the general population than among individual with DS (McDermott et al., 2005). However, it should be noted that the rate of seizure activity

among individuals with DS is lower than the reported rate among individuals with ID in general (McDermott et al., 2005). More recent estimates of the rate of seizures among adults with DS range from 9.4% to 26.5%, with a mean onset of seizures over the age of 30, around age 37 (Johannsen et al., 1996; McDermott et al., 2005; McVicker et al., 1994; Puri, Ho & Singh, 2001).

In addition to increasing with age, the rate of epilepsy and seizures is also related to the onset of dementia among adults with DS (Puri et al., 2001; Prasher & Corbett, 1993). In one study, 80% of the adults with seizures also presented with symptoms consistent with a clinical diagnosis of dementia (McVicker et al., 1994). In another study, 53% of individuals with dementia were reported to have seizures and the onset of seizures appeared to presage the onset of cognitive deterioration and symptoms of dementia (Lott & Lai, 1982). Indeed, the onset of seizures has been reported to occur at a younger age for individuals with DS who do not suffer from dementia (age 29) as compared to those who do (age 45). Dementia may be an important risk factor for late-onset seizures in adults with DS, but not for the high rate of seizures among all individuals with DS (Menéndez, 2005).

Thyroid dysfunction

Comparable to how the rate of seizures increases with age in adults with DS, the rate of individuals with DS at risk for thyroid disease also increases with age (Korsager, Chatham & Ostergaard-Kristensen, 1978), although some samples have indicated no increase with age (Murdoch, Ratcliffe, McLarty, Rodger & Ratcliffe, 1977; Šare, Ruvalcaba & Kelley, 1978). Approximately 35-40% of adults with DS are reported to have abnormal thyroid function, although only 7-8% had active hypothyroidism (Dinani & Carpenter, 1990; Prasher, 1994a; Prasher, 1994b). Also comparable to seizure findings, the rate of thyroid disease in adults with DS is greater than that found in the general population (Coleman, 1994).

The high rate of abnormal thyroid function highlights the need for consistent and routine monitoring of thyroid functioning for adults with DS. Routine monitoring is particularly important as some individuals tested with abnormal thyroid functioning were identified with thyroid disease and were receiving too much or too little medication to manage their disease (Prasher, 1994a).

Other Medical Conditions

Diabetes

There are a few reports of an increased risk of Type 1 diabetes among individuals with DS and of increased risk of mortality due to Type 1 diabetes as compared to the general population (Anwar, Walker & Frier, 1998; Hill et al., 2003). The age of onset of Type 1 diabetes appears to be increasing in individuals with DS. Earlier studies of Type 1 diabetes from the 1960s focused on children (Burch & Milunsky, 1969; Farquhar, 1962; Milunsky & Neurath, 1968) and reported that the age of onset peaked at 8 years of age for individuals with DS. However, the life expectancy of individuals with DS in the 1960s was 18 years (Collmann & Stoller, 1963). With increasing life expectancies for individuals with DS, more recent age of onset estimates of 22 years are comparable to the onset of type 1 diabetes in the general population (Anwar et al., 1998) and consistent with more individuals being diagnosed with adult-onset diabetes than juvenile diabetes. However, contrary reports suggest that adults with DS are at lower risk for mortality due to Type 1 diabetes than the general population and than adults with ID due to other causes (Haveman et al., 1989; Yang et al., 2002). Few reports are available regarding Type 2 diabetes in adults with DS, but the rate reported in a preliminary report appears to be lower than that in the general population

(Silverman, 2010). There is a need for more detailed population studies on the rate of Type 1 and Type 2 diabetes in individuals with DS to confirm these findings.

Obesity

A significant proportion of adults with DS are reported to be overweight or obese according to the body mass index (BMI). Between 45-79% of males and between 56-96% of females with DS are reported to be overweight (Bell & Bhate, 1992; Melville, Cooper, McGrother, Thorp & Collacott, 2005; Prasher, 1995; Rubin, Rimmer, Chicoine, Braddock & McGuire, 1998), while the prevalence in the general population that is overweight was reported to be approximately a third during the time period of these studies (Rubin et al., 1998). Contributing factors to the high rate of overweight and obese individuals with DS may include a combination of eating behavior, intake, metabolic rate, hypothyroidism and reduced exercise (Prasher, 1995).

BMI's are found to increase with age in the general population. Conversely, BMI's of cohorts of adults with DS are observed to be smaller with increasing age (Prasher, 1995; Rubin et al., 1998). Contributing factors to the lower BMI's found in older adults with DS are speculative, but may include residential setting, and healthier weights contributing to longevity (Rubin et al., 1998).

Sleep apnea

Obstructive sleep apnea (OSA) is present in approximately 30-55% of children with DS (de Miguel-Díez, Villa-Asensi & Álvarez-Sala, 2003; Stebbens, Dennis, Samuels, Croft & Southall, 1991). The known risk factors for OSA in children include facial (midfacial hypoplasia, mandibular hypoplasia) and other physical features (glossoptosis, an abnormally small upper airway, superficially positioned tonsils, relative tonsillar and adenoidal encroachment, hypotonia of upper airway) (Marcus, Keens, Bautista, von Pechmann & Ward, 1991; Roizen, 2003). Other risk factors have yet to be identified. Adults with DS are at increased risk for OSA as these risk factors observed in children continue into adulthood, and other risk factors become more prevalent. Obesity (reviewed previously) is a risk factor for OSA that put adults with DS at increased risk for developing this disorder. The rate of hypothyroidism (reviewed previously) increases with age in adults with DS, and is associated with OSA. The few studies that have examined the rate of OSA among adults with DS report that up to 94% of adults suffer from the disorder (Trois et al., 2009). Despite the increase in the number of risk factors predisposing adults with DS to OSA, increasing age is found to be the strongest risk factor, particularly when other risk factors are not present (Resta et al., 2003).

Musculoskeletal

Musculoskeletal problems often result from premature degenerative bone and joint disease (Dacre & Huskisson, 1988; Olive, Whitecloud & Bennet, 1988). Osteoporosis is common among adults with DS and adults are at greater risk as they age (Center, Beange & McElduff, 1998). There are several factors that may contribute to this increased risk for osteoporosis in adults with DS, including early menopause, decreased physical activity, low muscle tone and decreased strength. Degenerative osteoarthritis is also common among adults with DS, with osteoarthritis of the spine affecting 22% of middle-age adults and 40% of elderly adults (Van Allen et al., 1999). Typical symptoms include numbness, weakness and pain. It is often difficult to detect these symptoms in individuals with ID, and thus the rate of degenerative osteoarthritis may be underreported. Orthopedic problems, such as flat feet (a congenital condition), are the most common musculoskeletal abnormality observed in adults with DS, affecting approximately 70% of individuals (Prasher, 1994b).

Mitral valve prolapse

Mitral valve prolapse is common among adults with DS, occurring in between 46 to 57% of individuals (Barnhart & Connolly, 2007; Roizen & Patterson, 2003). Even though children with DS have a high risk of congenital heart disease, this previous cardiac pathology is not linked to the high rate of mitral valve prolapse later in life. Early signs of mitral valve prolapse are comparable to those observed in the general population and include fatigue, weight gain, and irritability (Barnhart & Connolly, 2007).

Mortality

Health problems present in adults, reviewed earlier in this chapter such as sensory handicaps, thyroid disorders, and degenerative spine disease, may contribute to the earlier mortality observed in adults with DS (Kapell, Nightingale, Rodriguez, Lee, Zigman & Schupf, 1998; Lantman-de Valk, Haveman, & Crebolder, 1996). However, their direct effect on mortality is not yet well documented. The life expectancy of adults with DS has a similar pattern until the age of 40 as compared to their age peers with other types of ID, and elevated mortality rates thereafter (Haveman, Maaskant & Sturmans, 1989; Maaskant, Gevers & Wierda, 2002; Strauss & Eyman, 1996).

The life expectancy of adults with DS is increasing and now averages around the mid- to late-50s. Still, this life expectancy is substantially below that of the general population and that of their peers with ID (Glasson, Sullivan, Hussain, Petterson, Montgomery & Bittles, 2002; Janicki, Dalton, Henderson & Davidson, 1999). Also, while women with DS are observed to have shorter life expectancies than men with DS, the opposite pattern is seen in the general population and among individuals with other types of ID (Carter & Jancar, 1983; Glasson et al., 2002; Glasson, Sullivan, Hussain, Petterson, Montgomery & Bittles, 2003; Tyrer, Smith & McGrother, 2007). A gender difference in mortality among individuals with DS is still speculative as other studies have not replicated this finding (Day, Strauss, Shavelle & Reynolds, 2005; Janicki et al., 1999). If valid, earlier menopause (discussed earlier) may be a contributing factor to earlier mortality in women with DS as compared to men with DS (Schupf, Zigman, Kapell, Lee, Kline & Levin, 1997).

The evidence is mixed regarding other risk factors found to predict mortality of adults with DS. Some studies have found that functional abilities predict mortality in adults with DS (Chaney & Eyman, 2000; Esbensen, Seltzer & Greenberg, 2007; Eyman, Call & White, 1991; Strauss & Zigman, 1996), comparable to findings in their peers with ID (Bittles, Petterson, Sullivan, Hussain, Glasson & Montgomery, 2002; Strauss & Eyman, 1996). Prior levels of functional abilities and declines in functional abilities were found to predict mortality in a sample of adults with and without DS (Esbensen et al., 2007). However, the relationship between functional abilities and mortality among adults with DS is not always supported (Glasson et al., 2002; Strauss & Eyman, 1996; Strauss & Zigman, 1996). Age has consistently been found to be a predictor of mortality for adults with DS (Esbensen et al., 2007; Eyman, Call & White, 1989; Hayden, 1998), and new research suggests that worsening of behavior problems is another predictor of mortality (Esbensen et al., 2007).

Common causes of death in this population include leukemia, respiratory illness, congenital circulatory defects, diseases of the digestive system, dementia and Alzheimer's disease, and are reported to vary with age (Day et al., 2005; Hermon, Alberman, Beral & Swerdlow, 2001; Hill et al., 2003; Thase, 1982). Although a common cause of death among children with DS is leukemia (behind respiratory illness and congenital heart defects), this risk is found to decrease with age (Hasle, Clemmensen & Mikkelsen, 2000; Yang et al., 2002). In contrast, the risk of mortality due to cancer in adults with DS is equal to or lower than that in the general population or among their peers with ID (Day et al., 2005; Hasle et al., 2000;

Patja, Eero & Iivanainen, 2001b; Sullivan, Hussain, Glasson & Bittles, 2007; Yang et al., 2002). In particular, the risk of mortality due to solid tumors among adults with DS is considerably lower than among their peers and the general population (Hasle et al., 2000; Hill et al., 2003; Sullivan, et al., 2007). In contrast, the risk for dementia was found to increase with age (Yang et al., 2002). Respiratory problems, congenital anomalies (other than congenital heart anomalies) and ischemic heart disease were also found to vary with age in their relation to mortality in individuals with DS. Among older adults with DS, respiratory problems and congenital anomalies are reported to be more common and ischemic heart disease less common than expected (Yang et al., 2002). That cardiovascular and circulatory defects are common causes of death among individuals with DS is not unexpected given the biological phenotype of this syndrome (Roizen, 1996). In comparison, the common causes of mortality found among individuals with ID include cardiovascular diseases, respiratory diseases, and cancers (Patja, Mölsä & Iivanainen, 2001a), and these have been found to be comparable to causes of mortality among a sample of only adults with DS (Esbensen et al., 2007).

Medical Conditions with Low Risk

While adults with DS are at risk for several medical conditions just described, they are also at low risk for other medical conditions. In a review of cancers among individuals with DS, malignant solid tumors were reported to be underrepresented (Satgé, Sommelet, Geneix, Nishi, Malet & Vekemans, 1998). In particular, common epithelial tumors were underrepresented in adults, as are breast, uterine, digestive, genital, skin, bronchial, ear/nose/throat or urinary tract cancers (Hasle, Clemmensen, Haunstrup & Margareta, 2000; Hill et al., 2003; Jancar & Jancar, 1976; Oster, Mikkelsen & Nielsen, 1975; Scholl, Stein & Hansen, 1982). However, a greater risk for testicular cancer has been reported among males with DS as compared to typically developing males (Dieckmann, Rube & Henke, 1997; Hasle et al., 2000).

Possible explanations for the decreased risk of some cancers among adults with DS are that accurate population morbidity studies are rare. It has also been proposed that individuals with DS may be less exposed to environmental contributors to cancer risk (Satgé et al., 1998). Decreased alcohol and tobacco use, early menopause and other lifestyle and environmental factors may contribute to lower cancer risk. However, obesity and the lack of physical activity common in adults with DS would contribute to an increased risk of cancers. The shorter life expectancy of individuals with DS has also been suggested to contribute to lower risk for cancer (Satgé et al., 1998). However, as the life expectancy of individuals with DS has increased dramatically over the last few decades, shorter life expectancy becomes a less probable explanation for the apparent lower risk of solid tumors. Several tumor-suppressor genes have been identified on chromosome 21, potentially contributing to the decreased risks for many solid tumors (Lee, T. Park, S. Park & J. Park, 2003). Copper-zinc superoxide dismutase, also located on chromosome 21, and its contribution to the metabolism of oxygen free radicals are further hypothesized to reduce the risk of carcinogens (de la Torre, Casado, Lopez-Fernández, Carrascosa, Ramirez & Saez, 1996).

While the rate of mitral valve prolapse is high, there is a lower risk for cardiovascular and cerebrovascular disease observed in adults with DS as compared to the general population (Marino & Pueschel, 1996) and lower rates of emphysema, fractures, hypercholesterolaemia and heart disease as compared to adults with ID due to other causes (Haveman et al., 1989; Kerins et al., 2008). Further, individuals with DS are found to have lower resting heart rates and lower blood pressure than the general population (Prasher, 1994b; Richards & Enver, 1979). The rise in blood pressure seen with age in the general population is not as great

among individuals with DS, and hypertension is an uncommon problem reported in adults with DS (Kerins et al., 2008; Prasher, 1994b).

While upper respiratory infections are common among adults with DS, significant respiratory problems are not common (Minihan & Dean, 1990; Prasher, 1994b; Wilson & Haire, 1990). Further, as mobility declines with age, recurrent pneumonia with incomplete recovery has been found to occur more often (Van Allen et al., 1999). This is particularly noteworthy as respiratory illness is a common cause of mortality in adults with DS. It may be that chronic respiratory problems contribute to mortality more so than acute respiratory problems.

Health Care

There are excellent guidelines for the health care of individuals with DS across the lifespan, including guidelines specific to adults (Cohen, 2002). They provide recommendations to health care professionals of what conditions to screen for and how frequently. However, little is known about the pattern of screening, health care use and access, or barriers to health care experienced by adults with DS specifically, although efforts are currently being made to explore these questions.

The literature on health care service use by adults with ID indicates that this population experiences significant health disparities in access to health care in comparison to the general population (Horwitz, Kerker, Owens & Zigler, 2000). As an example, few adults with ID receive care from specialists despite a high percentage of individuals, such as individuals with DS, having medical needs that require specialty care. An agenda has been set by the federal government for promoting the health of individuals with ID, improving their quality of and access to health care, and training health care providers to the specific needs of individuals with ID (US Public Health Service, 2001).

Conclusions

As individuals with DS continue to experience longer lives, the need to understand their aging and associated health conditions becomes more critical. The chronic disorders that onset in adults with DS, and the age-related change in other disorders, have important implications for health care management of this aging population. Health care providers need to be informed of the health conditions more common among adults with DS as they age, to be alert for declines earlier than expected in the general population, and the implication that early-life medical conditions may have in the later-life of the individual. For example, chronic and inadequately treated middle ear infections in childhood may have an impact on later hearing loss in adults with DS.

The different pattern of health conditions in aging adults with DS also has implications for family members and support providers. Over 60% of adults with ID co-reside with their families (Fujiura, 1998). As such, families and support providers of adults with DS need to be informed of what symptoms to be alert for in order to better communicate changes in health to medical providers, such as lethargy, irritability and fatigue. The health care provider is then responsible for determining whether these common symptoms are due to hypothyroidism, mitral valve prolapse, symptoms of menopause, pain, poor sleep, or to depression resulting from disorientation due to sensory impairments. Deteriorations in health can also be associated with an increase in behaviors, particularly if the individual with DS has communication difficulties in expressing problems or medical complaints. And finally, adults with DS should be provided with appropriate information to better understand, and counseling to cope with, changes in their own level of ability or health.

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