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ARTICLE

Health Care Concerns and Guidelines for Adults With Down Syndrome

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Down syndrome (DS) is the most common cause of mental retardation in North America, yet little information is available on the natural history of DS in adults. We report on significant medical problems of adults with DS (DS adults) residing in a British Columbia provincial residential center, Woodlands, over the 12-year period from 1981 through 1992. Prospective, yearly health care reviews on 38 DS adults are summarized according to age. Group 1 consists of 18 middle-aged DS adults less than 50 years old, and group 2 comprises 20 elderly DS adults 50 years and older. Significant health problems in all DS adults include untreated congenital heart anomalies (15.8%), acquired cardiac disease (15.8%), pulmonary hypertension (7.8%), recurrent respiratory infections/aspiration leading to chronic pulmonary interstitial changes (30%), complications from presenile dementia/Alzheimer-type disease (42%), adult-onset epilepsy (36.8%), osteoarthritic degeneration of the spine (31.6%), osteoporosis with resultant fractures of the long bones (55%) or vertebral bodies (30%), and untreated atlantooccipital instability (7.9%). Acquired sensory deficits are significant problems including loss of vision due to early onset of adult cataracts (50%), recurrent keratitis (21%) or keratoconus (15.8%), and significant hearing loss (25%). Behavioral problems (50%), loss of cognitive abilities, and onset of symptoms of Alzheimer disease (group 1: 5.5%; group 2: 75%) pose ongoing challenges for care. In conclusion, the quality of life for adults with DS can be improved by routine, systematic health care screening to identify treatable diseases that may be missed because of poor communication or confusion due to Alzheimer disease. Am. J. Med. Gent. (Semin Med. Genet.) 89:100–110, 1999. © **1999 Wiley-Liss, Inc.**

KEY WORDS: Down syndrome; adults with Down syndrome; natural history of Down syndrome; health-care screening

INTRODUCTION

Down syndrome (DS) is the most common cause of mental retardation in North America. A recent life-table study in British Columbia of live-born individuals with DS reports that over half survive into their fifties, 40% to age 60, and 13% to age 68 years [Baird and Sadovnick, 1987, 1988, 1989]. Improving societal acceptance and the creation of community programs and living settings for adults with DS (DS adults) are resulting in a shift in medical care from specialists working in residential centres to the community physician. Health problems of noncommunicative adults with intellectual impairment often go undetected, increasing the morbidity [Kerr et al., 1996; Piachaud et al, 1998].

As a result of the influx of DS adults into their practices, primary care physicians are asking for more information from specialists in order to address the health care needs of this population [Howells, 1986; Wilson and Haire, 1990, Kerr et al., 1996; Piachaud et al., 1998]. Surprisingly little information is available on the chronology of health care concerns of DS in adulthood. In this report we review significant medical problems of DS adults residing in a residential centre, Woodlands, over a 12-year period. The purpose of this study is to obtain longitudinal outcome information in order to delineate the natural history of DS in adulthood. This information will assist family physicians, community health care workers, and families in anticipating health care concerns and planning to meet the needs of DS adults.

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DS adults in residential care may not be representative of all individuals with DS due to intrinsic biases in case ascertainment. The reported population may have more health care and be-

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havioural issues than adults with DS living in the community. Nonetheless, we consider the paucity of information in the medical literature and the ready access to reliable information from the experience at Woodlands warrants our study.

METHODS

We reviewed comprehensive yearly medical assessments and acute care notes from medical records on 38 individuals with DS residing at one of the provincial residential centres for individuals with mental retardation in British Columbia, Woodlands. There was complete ascertainment of medical charts from all individuals with DS residing at Woodlands during the time period 1981–1992. Charts were divided into two groups. Group 1 included middle-aged DS adults less than 50 years old (reviewed by J.F.), and group 2 included elderly DS adults 50 years and older (reviewed by S.J.).

RESULTS

Demographics

In group 1, there were 18 middle-aged DS adults, 15 men and 3 women (Table I). At the time of chart review or death, their ages ranged from 30 to 43 years (mean age, 36.2 years). In group 2, there were 20 elderly DS adults, 13 men and 7 women. Their ages at the time of chart review, or death, ranged from 50 to 68 years old (mean age, 59 years).

Age at Admission

The age of admission was significantly different between the two groups. DS adults born between 1949 and 1963 were frequently admitted in early childhood, at a mean age of 7.3 years (range, 2 months to 13 years). Their mean length of residence at Woodlands was 31 years (range, 20–41 years). The average age of admission of elderly DS adults (born between 1917 and 1940) was 29.5 years (range, 6–54 years). Their mean length of residence at Woodlands was 28.25 years (range, 2–58 years). The discrepancy between groups in admission age reflects changing societal views and medical values as well as availability of accommodations in Woodlands.

Chromosome Diagnosis

Chromosome diagnosis was trisomy 21 in 34/38 and mosaicism for trisomy 21 in 2/38. No chromosome report was available for two others, but clinically the diagnosis of DS was not in question.

Deceased DS Adults

Thirteen of 38 of our DS adults were deceased. The mean age of death was 48.5 years (range, 32–67 years). In the middle-aged group, 4/18 were deceased. Lack of provision of cardiac surgical treatment, in keeping with health care policies at the time, resulted in the deaths of three of the four. Two individuals died at ages 32 and 36 years, respectively, from complications of uncorrected atrioventricular canal defects and pulmonary hypertension. One man died at age 35 years from complications of rheumatic heart disease with valvular changes causing aortic insufficiency and mitral valve stenosis. The fourth man died unexpectedly at 36 years old from bilateral pneumonia.

In the elderly group, 9/20 were deceased. At the time of death, 7/9 had a clinical diagnosis of advanced Alzheimer disease; they were bedridden and noncommunicative, requiring 24-hour nursing care for assistance with all their needs. Autopsies done on 3/9 (who died at ages 53, 61, and 65 years, respectively) confirmed advanced Alzheimer disease changes in the brain, terminal pneumonia, and arteriosclerosis. No autopsies were done on the remaining 4/9 who had advanced Alzheimer-type disease. Their ages ranged from 63 to 67 years. Before death, all were bedridden for a number of years, were minimally responsive, and had clinical evidence of Alzheimer-type disease. Death was attributable to pneumonia in all four.

Two of the nine who were deceased had clinical evidence of early Alzheimer-type disease. One man had post-traumatic paraplegia and died of pneumonia at 54 years old following an orthopaedic operation. The other man died unexpectedly at 48 years old during an influenza epidemic. He had an adult-onset seizure disorder. There was inadequate information to assess the clinical question of whether the seizure

TABLE I. Demographic Information on Adults With Down Syndrome (DS-A) Residing at Woodlands

Demographic feature	Middle aged DS-A (n = 18)	Elderly DS-A (n = 20)
Birth years	1949–1963	1917–1940
Age at time of chart review	36.2 (30–43) years old	59.7 (47–68) years old
Age at admission	7.3 years old (2 months–13 years old)	32.5 (6–54) years old
Length of stay	31 (20–41) years	25.8 (1–49) years
Sex	15 men and 3 women	13 men and 7 women
Deceased by 1992	4/18	9/20
Severe to profound mental retardation	18/18	20/20
Karyotypes	16/18 47,N,+21	18/20 47,N,+21
	2/18 47,N,+21/46,N	2/20 no karyotype

disorder stemmed from early symptoms of Alzheimer-type disease.

Cognitive Deficits

All DS adults in this study had developmental testing either before admission or shortly after admission. All were assessed to have severe to profound mental retardation. It was not possible to assess loss of cognitive function over time because this information was not in the medical charts. There was comment in the yearly medical reviews regarding abilities in activities of daily living and behavioural concerns. This information was reviewed for middle-aged DS adults (Table II).

Health Care Concerns

Frequently encountered and significant health concerns are summarised by organ system in Table II.

DISCUSSION

Multiple health concerns are common in DS adults residing in Woodlands. Whether DS adults are living in the community or in residential care, a program of anticipatory health care screening and early detection of health problems is recommended (Table III). Rigorous, systematic screening of adults with DS, and other intellectual disabilities with communicative disorders, may minimize the morbidity and long-term complications, thus improving the quality of life.

The age of the individual, their birth year, age at time of admission, and the reason for admission are all contributing factors to the types of health care concerns. At the time of admissions, those born between 1949 and 1963 are younger (mean age, 7.3 years) and healthier than those born before 1945. Almost all DS adults (92%, 36/38) have admissions to Woodlands between 1945 and 1980. This admission pattern reflects the social and political milieu in British Columbia.

During the time period evaluated in this study, Woodlands was considered to be the premiere residential school and training centre for individuals with intellectual disabilities. Community schools did not have programs for the disabled; for this reason, "enlightened" parents sought out admission to Woodlands so that their DS children could be educated and trained in an occupation [Adolph, 1996]. Woodlands also provided 24-hour nursing care and rehabilitation programs for disabled individuals with significant health concerns, such as congenital heart anomalies. Community residential centers in homes and integrated education programs in schools became more prevalent in British Columbia in the 1970s. After 1980, a deliberate effort was made to place all the 1,400 disabled persons residing at Woodlands in community settings. Woodlands has closed, except for the Willows clinic for multidisciplinary assessment of health, psychiatric, and behavioural concerns and for residential care of dually diagnosed individuals who cannot be placed in community settings.

Whether adults with Down syndrome are living in the community or in residential care, a program of anticipatory health care screening and early detection of health problems is recommended.

Those medical problems that pose the greatest health risks to DS adults include pulmonary hypertension associated with untreated congenital heart anomalies, acquired cardiac diseases, recurrent respiratory infections and aspiration, complications attributable to presenile dementia/Alzheimer-type disease, adult-onset epilepsy, osteoarthritic degeneration of the spine, osteoporosis and resultant fractures of the long bones and vertebral bodies, and untreated atlantooccipital instability, hypothyroidism, and arthritis. Acquired sensory deficits pose significant problems with respect to both delayed diagnosis and long-term morbidity, resulting in decreased enjoyment of daily activities. Of particular concern are loss of vision due to early onset of cataracts, recurrent keratitis and keratoconus, and deafness resulting from chronic middle ear infections and presbycusis. Behavioural problems related to the underlying cognitive deficits, loss of abilities with advancing age, and onset of symptoms of Alzheimer disease pose ongoing challenges in everyday care of DS adults.

Eye Disorders

Previous reports confirm the high incidence of eye abnormalities in DS adults [Hestnes et al., 1991]. Normal ocular findings in DS adults are considered to be an exception, and the possibility of ocular pathologic conditions should be actively investigated. Diagnosis of early cataracts and keratoconus may be difficult in marginally cooperative patients. In childhood, the chief concerns for DS individuals are congenital cataracts, refractive errors, strabismus, and amblyopia. With increasing age, adult-onset cataracts are common [Pueschel, 1990; Evenhuis, 1991]. Among the Woodlands patients, refractive errors (32.4%, 13/38), usually myopia, and strabismus (36.8%, 14/38) are common. Adult-onset cataracts are present in 33% of our middle age and in 65% of our elderly DS adults. Even though cataract removal is possible, visual problems related to aphakia and poor compliance with use of bifocals result in significant visual disability even in those who are treated.

Keratoconus is another condition frequently reported in adults with DS. The pathogenesis of this condition is not known. Hestnes et al. [1991] reports on 9 DS patients with keratoconus, two of whom developed phthisis bulbi. Keratoconus is more common in our elderly (20%) than in our middle-aged (11%) DS adults. Only one of our adults with keratoconus developed phthisis bulbi. Frantz et al. [1990] reports favourable results of keratoplasty in individuals with DS. Careful selection of patients and individual nursing during the postoperative period are recommended.

TABLE II. Health Concerns in Adults With Down Syndrome (DS-A)

	Middle-aged DS-A (%)	Elderly DS-A (%)	All DS-A (%)
Visual concerns			
Congenital cataracts	1/18	0/20	1/38 (2.6)
Adult-onset cataracts	6/18	13/20	19/38 (50)
Blindness	3/18	5/20	8/38 (21)
Strabismus	6/18	8/20	14/38 (36.8)
Recurrent keratitis	3/18	5/20	8/38 (21)
Keratoconus	2/18	4/20	6/38 (15.8)
Refractive errors	4/18	9/20	13/38 (34.2)
Total	15/18 (39.5)	15/20 (75)	30/38 (79)
Hearing concerns			
Chronic otitis media	5/18	6/20	17/38 (44.7)
Hearing loss	NI	5/20	5/20 (25)
Chronic mastoiditis	3/18	4/20	7/38 (18.4)
Mastoidectomy	2/18	4/20	6/38 (15.8)
Total	10/18 (26.3)	7/20 (35)	17/38 (44.7)
Respiratory concerns			
Pulmonary hypertension	2/18	1/20	3/38 (7.9)
Pneumonia	9/18	12/20	21/38 (55.3)
Asthma/bronchitis	1/18	2/20	3/38 (7.9)
Recurrent aspiration with chronic interstitial lung changes on X-ray	NI	6/20	6/20 (30)
Total	11/18 (29)	12/20 (60)	23/38 (60.5)
Cardiovascular concerns			
CHD	3/18	3/20	6/38 (15.8)
Ventricular septal defect	1/3	2/3	3/6
Atrioventricular canal defect	2/3	0	2/6
Patent ductus arteriosus	0	1/3	1/6
Rheumatic heart disease	1/18	0/20	1/38 (2.6)
Arteriosclerotic heart disease	1/18	4/20	5/38 (13.2)
Cerebral vascular accidents	0/18	2/20	2/38 (5.3)
Hypertension	0/18	0/20	0/38 (0)
Total	11/18 (61)	11/20 (55)	22/38 (57.9)
Gastrointestinal concerns			
Hiatal hernia	0/18	3/20	3/38 (7.9)
Gastroesophageal reflux	1/18	6/20	7/38 (18.4)
Fundoplication for gastroesophageal reflux	1/18	0/20	1/38 (2.6)
Prolapsed rectum	0/18	3/20	3/38 (7.9)
Hernias			
Ventral	NI	2/20	2/20
Incisional	NI	1/20	1/20
Inguinal	NI	1/20	1/20
Total	11/18 (61.1)	5/20 (25)	6/38 (15.8)
Genitourinary concerns			
Recurrent urinary tract infection	0/18	3/20	3/38 (7.9)
Bladder neck resection	0/18	4/20	4/38 (10.5)
Hydronephrosis	0/18	2/20	2/38 (5.2)
Renal failure	0/18	1/20	1/38 (2.6)
Total	4/18	8/20	12/38 (31.6)
Musculoskeletal			
Paraplegia due to compression fracture	1/18	1/20	2/38 (5.3)

(*Continued*)

TABLE II. Health Concerns in Adults With Down Syndrome (DS-A) (Continued)

	Middle-aged DS-A (%)	Elderly DS-A (%)	All DS-A (%)
Quadraplegia due to atlantooccipital instability	1/18	0/20	1/38 (2.6)
Compression fracture of vertebral body with osteoporosis	0/18	6/20	6/38 (15.8)
Osteoarthritis			
Spine	4/18	8/20	12/38 (31.6)
Other joints	0/20	5/20	5/38 (13)
Atlantooccipital instability	1/18	2/20	3/38 (7.9)
Fractures			
Long bone	NI	11/20	17/20 (85)
Other bones	NI	6/20	6/20 (30)
Vertebral bodies	0/18	6/20	6/20 (30)
Total	5/18 (27.7)	17/20 (85)	22/38 (57.9)
Central nervous system			
Seizures	9/18	11/20	20/38 (52.6)
Childhood onset	3/18	0/20	3/38 (7.9)
Adult onset	6/18	11/20	17/38 (44.7)
Alzheimer disease	1/18	15/20	16/38 (42)
Parkinsonian symptoms	0/20	1/20	1/38 (2.6)
Behavioural problems	12/18	7/20	19/38 (50)
Self-abusive	9/18	NI	9/18 (50)
Aggressive	5/18	NI	5/18 (27.7)
Passive	5/18	NI	5/18 (27.7)
Autistic-like	3/18	NI	3/18 (7.9)
Total	9/18 (50)	11/20 (55)	20/38 (52.6)
Other health concerns			
Hypothyroidism	4/18	7/20	11/38 (28.9)
Vitiligo with alopecia areata	2/18	0/20	2/38 (5.3)
Trichophyton rubrum	1/18	0/20	1/38 (2.6)
Discoid lupus erythematosus	1/18	0/20	1/38 (2.6)
Chronic idiopathic thrombocytopenic purpura	0/18	1/20	1/38 (2.6)
Hepatitis B carrier	8/18	2/20	10/38 (26.3)

Hearing Disorders

Recurrent and chronic ear infections in noncommunicative adults often go undiagnosed and can lead to mastoiditis and hearing loss. Almost half of the DS adults in our study have hearing concerns, most frequently secondary to recurrent ear infections. 18% have a history of mastoiditis, usually resulting in mastoidectomy. Significant hearing loss is present in at least 25% of our elderly group. Difficulties in testing and inconsistent reporting in yearly medical reviews underestimates the presence of less severe hearing losses. Balkany et al. [1979] report on significant hearing loss in 78% of 107 individuals with DS, age 2 months to 60 years, tested with otoscopy, audiometry, and impedance tympanometry. The most complete study available on hearing concerns among DS adults living in residential centres is reported by Evenhuis et al. [1991]. Thirty-five adults, age 35–62 years, are evaluated with otoscopy, pure tone audiometry, impedance audiometry, and auditory evoked responses. A loss of hearing greater than 20 dB is present in 59 ears tested. The documented hearing losses are conductive, presumably resulting from middle ear disease, and sensorineural.

Cardiovascular Diseases

Congenital heart anomalies (CHA) are a common structural anomaly in DS, occurring in 42% of newborn infants evaluated by echocardiography [Tubman et al., 1991]. The incidence of CHA in surviving adults is unknown. In the Woodlands population, 15.8% of DS adults have untreated CHA including three with ventricular septal defects, two with atrioventricular canal defects, and one with a patent ductus arteriosus.

An additional 10 of 38 DS adults in our study have significant cardiac murmurs without cardiac symptoms. Hamada et al. [1998] report a high incidence of valvular heart abnormalities in 30 institutionalised DS adults with no history of congenital heart anomalies and no cardiac symptoms. Echocardiograms on these adults show that 26.7% have mitral valve prolapse, with an increase in 20% of echo brightness of the mitral valve; 16.75% have mitral valve regurgitation; and 13.3% have aortic valve regurgitation. Rheumatic heart

TABLE III. Health Care Screening of Adults With Down Syndrome[†]

Health assessment	Initial	Annual	Periodic	PRN
Central and peripheral nervous system				
Neurological history and exam	X	X		X
Cognitive skills assessment	X	X	q 5 yrs	X
Behavioural assessment	X	X		X
Seizure evaluation	X	X		X
EEG	⋆	⋆		X
Neurology consultation	⋆⋆	⋆·⋆⋆		X
Ear, Nose, and Throat (ENT)				
History and exam	X	X		X
ENT specialist consultation	⋆⋆	⋆·⋆⋆		⋆⋆
Visual system				
History and exam	X	X		X
Ophthalmology consultation	X	⋆·⋆⋆		⋆⋆
Auditory system				
History and exam	X	X		X
Hearing testing	X	⋆	q 3–5 yrs	X
Audiology consultation	⋆⋆	⋆·⋆⋆		⋆⋆
Pulmonary system				
History and exam	X	X		X
CXR	X	⋆		X
Tuberculosis skin test	X		q 5 yrs	
Specialist consultation	⋆⋆	⋆·⋆⋆		⋆⋆
Haematological system				
History and exam	X	X		X
Complete blood count, differential, platelets	X	X		X
Haematology consultation	⋆⋆	⋆		⋆⋆
Cardiovascular system				
History and exam	X	X		X
CXR/echocardiogram	X	⋆		X
Lipid profile	X	⋆		
Cardiology consultation	⋆·⋆⋆	⋆·⋆⋆		⋆⋆
Gastrointestinal system				
History and exam	X	X		X
Hepatitis screening/immunisation	X			
Specialist consultation	⋆·⋆⋆	⋆·⋆⋆		⋆⋆
Genitourinary system				
History and exam	X	X		X
Sexual activity/knowledge counselling	X	+	+	
Urinalysis, renal function tests	X	X		X
Routine pelvic exam and PAP smear	X	+	+	
Specialty consultation	⋆·⋆⋆	⋆·⋆⋆		⋆⋆
Endocrine and autoimmune disorders				
History and exam	X	X		X
Thyroid function tests	X	X		X
Diet counselling	X	X		
Specialty consultation	⋆·⋆⋆	⋆·⋆⋆		⋆⋆
Musculoskeletal system				
History and exam[a]	X	X		X
Spine radiography/odontoid views	X	X		
Counsel regarding sports activities	X	X		
Orthpaedic consultation	⋆·⋆⋆	⋆·⋆⋆		⋆⋆

[†]q 5 yrs, every 5 years formal testing with specialist; X, evaluate focussing on concerns related to DS; ⋆, evaluate based on clinical assessment and/or specialist's recommendation; ⋆⋆, obtain consultation if there is history of concerns, ongoing concerns, or suspected new concerns; +, screening frequency based on individual DS adults, their tolerance, and activities.

[a]Assess for osteoporosis.

disease is documented in only one man, who died of aortic insufficiency and congestive heart failure at age 35 years. Certainly, further diagnostic investigation is warranted on DS adults who develop heart murmurs.

Three of 38 DS adults have documented myocardial infarctions—a 65-year-old woman, a 61-year-old man, and a 35-year-old man with a history of a nonhypertensive stroke. Arteriosclerotic changes are reported in 6/8 deceased DS adults with autopsies. Routine investigation for lipid disorders is not part of the yearly assessment at Woodlands. Hypertension is not a finding in any of the assessed individuals in our study.

Respiratory Tract Diseases

Both acute and chronic problems of the respiratory tract occur frequently in our population. Three of the individuals with documented congenital heart anomalies also have pulmonary hypertension. Pulmonary hypertension, which is earlier in onset and often the cause of death in untreated DS individuals, is well reported in the literature. Other pulmonary problems are common, including documented pneumonia in 55.2% of DS adults at some time during their residence at Woodlands. Recurrent pneumonia with incomplete recovery occurs more often as mobility declines. Of grave concern are six elderly DS individuals with chronic interstitial changes of the lungs (documented by radiography) of insidious onset, attributable to chronic, recurrent aspiration. Respiratory problems related to aspiration are known to be associated with lower oesophageal sphincter incompetence and gastroesophageal reflux disease in the general population in patients with neurogenic dysphagia, obesity, and a sedentary lifestyle; and in handicapped persons without overt neurological deficits [Sulkes, 1986].

Gastrointestinal Disorders

The most common gastrointestinal problems in infants with DS—duodenal atresia and Hirschprung disease—are not reported in our adult population. Although aspiration pneumonitis and chronic interstitial changes from presumed gastroesophageal reflux are apparent, only one of our DS adults is diagnosed with gastrointestinal reflux, recurrent esophagitis, and stricture requiring surgical treatment. Hiatal, ventral, postoperative incisional, and inguinal hernias are found in 20% of elderly DS adults. The hernias may be related to connective tissue laxity, which occurs in DS individuals of all ages.

Obesity is a common concern among DS individuals, occurring in 70% of men and 95% of women [Bell and Bhate, 1992] compared with 50% of individuals with other forms of mental disability. Obesity is common among DS individuals admitted to Woodlands as adults (10/13) and appears to improve with dietary restrictions. Obesity resulting in hypoventilation syndrome is documented in one resident, presenting clinically as hypersomnia. Normal weight tables for adults with DS are not available. The basal metabolic rate in healthy DS individuals is comparable to that of normal controls [Schapiro and Rapoport, 1989], implying that dietary interventions can be effective.

Constipation is a frequent complaint, particularly among the inactive elderly DS adults. Rectal prolapse is reported in 3/8 of our adults with chronic constipation. There is controversy in the medical literature concerning whether or not celiac disease occurs more frequently in DS individuals compared with the general population [Tolmie, 1996]. None of our adults have a diagnosis of celiac disease.

Urinary Tract Disorders

Recurrent urinary tract infections (UTIs) are of particular concern in our population. UTIs were documented in more than 25% of DS adults, particularly those who are bedridden. Bladder neck resection is reported in 4/20 of the older DS adults and none of the younger group. Mesangiocapillary glomerulonephritis, focal segmental glomerulosclerosis with hylanosis, acute glomerulosclerosis, minimal change disease, and membranous nephropathy causing renal failure are reported as occurring in DS [Gupta et al., 1991; Lo et al., 1998].

Among our adults, renal failure is present in one. This is attributed to chronic, recurrent pyelonephritis. Another man, a carrier of hepatitis B, has persistent proteinuria and haematuria with normal blood pressure. His clinical picture is attributed to minimal glomerular change disease associated with being a hepatitis B carrier. Hydronephrosis is documented in one patient. Other structural renal anomalies are not reported in our population; hoever, systematic renal ultrasound studies are not part of the routine yearly medical evaluations. It is recommended that DS adults be assessed for structural renal anomalies with abdominal sonograms, along with routine monitoring for renal, and especially glomerular, disease [Lo et al., 1998].

Musculoskeletal Disorders

Premature degenerative bone and joint disease in adults with DS is reported in the orthopaedic literature [Dacre and Huskisson, 1988; Olive et al., 1988]. Pain and limitation of movement related to degenerative osteoarthritis is a common concern in our population. Among the middle-aged DS adults, osteoarthritis of the spine is reported in 22% (4/18); none have osteoarthritis of other joints. In the elderly group, 40% (8/20) have osteoarthritis of the spine with five of these eight having osteoarthritis in other joints. The radiological presentation of spinal osteoarthritis in DS adults is similar to that reported in other geriatric patients, although the symptoms appear to occur at a younger age among patients with DS [Tangerud et al., 1990]. Joint laxity is the attributable cause for early degenerative bone disease, but other factors may also be important.

Common symptoms related to spinal degeneration—paraesthesias, numbness, weakness, and pain—may go unreported in less communicative adults with DS [Voskuhl and Hinton, 1990]. Carpal tunnel disease occurs frequently among DS adults and needs to be con-

sidered in the differential diagnosis [Christensen et al., 1998]. To detect spinal degeneration, yearly clinical evaluations require careful neurological examinations assessing for changes in sensation, ambulation, continence, spine mobility, selective muscle atrophy, as well as deep-tendon reflexes. Early loss of ambulation among elderly DS adults is attributable to osteoarthritis as well as Alzheimer disease. Diagnostic investigations are indicated, since symptoms related to osteoarthritis are treatable [Taylor et al., 1991].

Fractures that are secondary to osteoporosis and trauma are common in our population. In the elderly group, 11/20 have a history of long-bone fractures, 6/20 have fractures of other bones, and 6/20 have documented collapse of vertebral bodies. Paraplegia stemming from a compression fracture of the spine is reported in one middle-aged resident who was injured after jumping out a window. Another elderly DS man has a compression fracture due to severe osteoporosis. Decreased exercise, inadequate dietary calcium, and limited exposure to sunlight in the Vancouver climate are all contributing factors to increased osteoporosis in our population. Other metabolic factors related to the underlying aneuploidy may also have contribute to this clinical problem.

Atlantoaxial subluxation is a serious but infrequent complication of DS [Pueschel and Scola, 1987; Davidson, 1988; Morton et al., 1995; Tolmie, 1996]. Reports suggest that the radiological gap decreases with age, presumably as laxity of the ligaments decreases, reducing the risk of spinal cord trauma [Tolmie, 1996]. One of our middle-aged adults has paraplegia from atlanto-occipital instability. Two elderly adults have atlantooccipital instability without neurological symptoms.

Cancer

Cancers, including leukaemias, are not reported in any of our adults. Baird and Sadovnick [1990] report on the attributable causes of death from the certificate data of 324 individuals with DS, nine of whom died of unspecified tumors. Overall, the rate of leukaemia in DS is estimated to be one in 150, which represents a 20-fold increased risk over the general population risk [Zipursky et al., 1992]. This risk increase is thought to extend into adulthood. Testicular germ cell tumours may be more frequent in adult men with DS compared with the general population [Dieckmann et al., 1997]. Satge et al. [1998] review cancers in people with DS, finding that most malignant solid tumours occur less frequently in DS compared to the general population. The solid tumours that Satge et al. [1998] identifies as being more common in DS are lymphomas, extragonadal germ cell tumours, testicular germ cell tumours, and, a possible increase in retinoblastoma and pancreatic and bone tumours. "Lifestyle" cancer risks, such as for lung and cervical cancers, appear to be lower among those with DS.

It is unclear whether screening for cancer in DS adults should be more comprehensive than the yearly physical examination, routine lab studies, and symptom-related investigations, in particular, when testing is painful and requires dutiful cooperation. Pap smears, screening colonoscopy, mammograms, and other similar investigations can be traumatic experiences for noncommunicative adults with DS and other disorders associated with intellectual disabilities. The yield of these investigations needs to be balanced against the actual risks and other overriding health issues.

Central Nervous System Concerns

Epilepsy

The prevalence of seizures in DS appears to peak in infancy and late in life. Seizures occur in 5–10% of children with DS [Pueschel et al., 1991; Stafstrom et al., 1991; McVicker et al., 1994]. Different seizure types are reported, including infantile spasms, tonic–clonic, myoclonic, and generalized seizures. In our population, seizures occur in 50% (9/18) of middle-aged and 55% (11/20) of elderly DS adults. Three have a history of onset of seizures in childhood, one of whom is reported to have had infantile spasms. All others have onset of seizures in adulthood. Seizures can be associated with the onset and progression of Alzheimer-type changes in the brain [Stafstrom et al., 1991], being largely responsible for the increase in epilepsy among elderly patients with DS.

Alzheimer disease

The literature states that "almost all" DS adults 40 years and older have evidence of declining abilities and evidence of Alzheimer disease changes on magnetic resonance imaging (MRI), EEG, and neuropathological examination of the brain. Clinical dementia stemming from Alzheimer disease appears later in the coure of the illness. In a study of 53 individuals with DS living in institutions [Lai and Williams, 1989], dementia was found to be present in 8% (2/25) of patients 35–49 years old, 55% (11/20) of patients between 50 and 59 years old, and 75% (6/8) of those patients over 60 years old. Eighty-four percent of the reported demented patients have seizures, and 20% have symptoms of Parkinson disease. DS men are three times more likely to develop Alzheimer disease symptoms compared with DS women [Schupf et al., 1998]. DS adults with apolipoprotein E3/4 and 4/4 genotypes are four times more likely to develop Alzheimer disease symptoms compared to those with the 3/3 genotype [Schupf et al., 1998].

In our population, evidence of dementia is present in one (1/18) of the middle-aged group (30–43 years old) and in 75% (15/20) of the elderly group. Stratifying the elderly, DS individuals, onset of dementia is present in 66% (8/12) of those between 43 and 60 years old and 87.5% (7/8) of those over 60 years old. One of our elderly DS individuals had parkinsonian symptoms associated with Alzheimer disease. Visser et al. [1997] report on prospective evaluations of clinical signs, cognitive functioning, and EEG results in 307 adults with DS living in institutions. Progressive mental and physical deterioration is found in 56 of the 307, with a mean age of onset of dementia at 56

years. The prevalence is 11% among 40- to 49-year-old adults, rising to 77% in those who are 60–69 years old.

MRI findings in DS adults and dementia demonstrate reduced total brain mass, particularly in the hippocampal and amygdala regions, and increases in ventricular size [Pearlson et al., 1998]. Hippocampal size is also reduced in normal DS adults, so that reduction in the size of the amygdala is a more specific sign of dementia [Aylward et al., 1999]. Prasher et al. [1996] suggest using MRI as a tool for diagnosis of Alzheimer disease in DS adults, particularly when standardised intellectual assessments are not possible.

Even without dementia, adults with DS are known to have declining cognitive function [Haxby, 1989; Caltagirone et al., 1990; Das et al., 1995]. Depression [Cooper and Collacot, 1994; Prasher and Hall, 1996] and other psychiatric disorders [Myers and Pueschel, 1991] can also interfere with their ability to carry out normal tasks of daily living.

Declining cognitive function in DS adults is observed in our population and is reported by others [Francheschi et al., 1990; Pary, 1992; Politoff et al., 1996]. For all 13 adults with DS admitted to Woodlands in adulthood, the reason for admission is declining abilities and behavioural problems limiting their ability to remain in a home setting. Significant factors contributing to decline in adaptive behaviour are ageing, dementia, and severity of mental retardation [Schapiro et al., 1992; Prasher and Chung, 1996] as well as major illnesses. Absence of medical illness is a positive predictor of a higher level of adaptive behaviour in elderly DS adults [Prasher and Chung, 1996].

The previous lack of objective and quantitative assessment tools for individuals with moderate to profound mental retardation limits our ability to document cognitive changes in the Woodlands DS adults. Witts and Elders [1998] reports on the reliability of the "Severe Impairment Battery" for assessing DS adults. Das et al. [1995] confirm that older individuals with DS perform more poorly when assessed by the Das-Haglieri cognitive assessment system and other tests of intellectual functioning. Use of one of these tools is recommended for periodically evaluating adults with DS in conjunction with routine health care screening.

Known confounding factors in diagnosing dementia in DS include depression, thyroid dysfunction, sensory deficits (e.g., progression of cataracts and hearing loss), neuroleptic-induced dementia [Gedye, 1998], and medication overdosage, particularly of anticonvulsants and antipsychotics. Although only one of our middle-aged DS individuals have a diagnosis of early dementia, the clinical notes on other individuals are suggestive of declining abilities. In several of the older individuals, stressful life events, such as an operation or a change in residence, precipitated a sudden and lasting global deterioration. One such DS individual died within a year of admission, following the death of his mother and relocation from living at home to residing at Woodlands.

Infections

Problems with cell-mediated and humoral immunity are well documented in DS individuals [reviewed by Tolmie, 1996]. Recurrent infections, particularly upper-respiratory-tract, middle ear, and skin infections, are common in the Woodlands population. It is difficult to determine whether the incidence of infections in DS individuals at Woodlands is increased compared to other residents, so it is not possible to comment further. Chichon et al. [1998] hypothesize that the increase in periodontitis associated with DS is secondary to an exaggerated immune inflammatory response.

The middle-aged and elderly Down syndrome individuals in this study have multi-system medical problems. Clinical symptoms and signs of early disease are frequently missed because of limited communicative skills.

Hepatitis B surface antigen is more prevalent among individuals with DS, independent of whether they are in institutional care or living with their families [Madden et al., 1976; Renner et al., 1985]. Hepatitis B antibodies are documented in eight of our middle-aged group and five of the elderly group. Ten of 13 are considered to be chronic hepatitis B carriers. Screening of Woodlands residents in 1985 identifies 7% of residents to be hepatitis B carriers. The majority of these carriers (20/38) have DS. This rate is far in excess of DS representation among Woodlands' residents (42/543–in 1985). Immunisation of nonimmune DS adults against hepatitis B, as well as hepatitis A and other common immunisable disorders, is strongly recommended.

Other Health Concerns

Thyroid disorders and other autoimmune disorders are common in individuals of all ages with DS [Loudon et al., 1985; Pueschel and Pezzullo, 1985; Karlsson et al., 1998]. In a survey of 106 adults with DS, more than 30% have elevated microsomal autoantibodies, but only 5% have hypothyroidism, and an additional 4% have elevated thyroid-stimulating hormone with normal T_3 and T_4 levels [Dinani and Carpenter, 1990]. A subsequent longitudinal study by Karlsson et al. [1998] of 85 individuals with DS followed up to age 25 years reports a much higher rate of thyroid abnormalities. Thirty five percent (30/85) have hypothyroidism, 2.3% (2/85) have hyperthyroidism. In the Woodlands population, hypothyroidism is being treated in 22.2% (4/18) of the middle-aged group and 35% (7/20) of the elderly DS individuals. Yearly screening with thyroid-stimulating hormone and T_4, along with a high index of suspicion, is recommended for early

detection [Murdoch et al., 1977; Tolmie, 1996].

One of our residents had discoid lupus erythematosus. Vitiligo and alopecia areata are also mentioned in the records of several patients. There is inconsistency in reporting these concerns, so that the frequencies cannot be adequately determined.

Health Screening

The middle-aged and elderly DS adults in this study have multisystem medical problems. Clinical symptoms and signs of early disease are frequently missed because of limited communicative skills. Routine, systematic screening for common health concerns in adults with DS is recommended. Table III summarises suggested guidelines for health care screening. At a minimum, the yearly physician evaluations can focus on the problems identified as common health concerns. Systematic health care screening can improve the quality of life of adults with DS through early detection of preventable disorders, particularly eye and ear infections. When there is loss of skills, investigations need to be initiated excluding treatable diseases, such as hypothyroidism, prior to assuming the symptoms herald the onset of Alzheimer disease. Education of guardians of DS adults with can also help in early identification and recognition of illnesses. The ultimate goal of health care screening is for DS adults to live enjoyable lives, free from undiagnosed and untreated chronic health concerns.

When there is loss of skills, investigations need to be initiated excluding treatable diseases, such as hypothyroidism, prior to assuming the symptoms herald the onset of Alzheimer's disease.

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