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EFFECTS OF AGE ON THE ADAPTIVE BEHAVIOR OF INSTITUTIONALIZED ADULTS WITH DOWN SYNDROME

BY

DIANNE E. RASMUSSEN 

A THESIS

SUBMITTED TO THE FACULTY OF GRADUATE STUDIES AND RESEARCH IN PARTIAL FULFILLMENT OF THE REQUIREMENTS FOR THE DEGREE OF MASTER OF EDUCATION

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THE UNDERSIGNED CERTIFY THAT THEY HAVE READ, AND RECOMMEND TO THE FACULTY OF GRADUATE STUDIES AND RESEARCH FOR ACCEPTANCE, A THESIS ENTITLED **EFFECTS OF AGE ON THE ADAPTIVE BEHAVIOR OF INSTITUTIONALIZED ADULTS WITH DOWN SYNDROME** SUBMITTED BY DIANNE E. RASMUSSEN IN PARTIAL FULFILLMENT OF THE REQUIREMENTS FOR THE DEGREE OF MASTER OF EDUCATION IN EDUCATIONAL PSYCHOLOGY.

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ABSTRACT

This study investigated age-related changes in adaptive functioning in institutionalized adults with Down syndrome as compared to non-Down syndrome mentally handicapped adults of the same sex and functioning level. From mentally handicapped residents of The Michener Centre, 140 subjects with Down syndrome were selected and matched on age, level of functioning, gender, and sensory handicap. Subjects were divided into two age groups, over or under 40 years of age, and comparisons were made across 20 adaptive skill subdomains. All had been assessed between 1985 and 1990 by Michener Centre Psychological Services staff on The Pyramid Scales: Criterion Referenced Measures of Adaptive Behavior in Severely Handicapped Persons (Cone, 1984).

Results obtained via cross-sectional methodology indicated that significant differences were related to level of functioning rather than to age or etiology of disability. Longitudinal analysis revealed that adaptive skills do show a greater decline with age in several Down syndrome persons beyond age 40. A pattern of decline in self-help and communication skills was identified for this group. Case studies of 4 Down syndrome individuals with confirmed Alzheimer pathology at postmortem were presented.

Results of the study are discussed in relation to aging, and the likelihood of Alzheimer-like changes in Down syndrome persons.

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TABLE OF CONTENTS

LIST OF TABLES

LIST OF FIGURES

CHAPTER I

Introduction and Statement of the Problem

Down syndrome is the most common cause of mental retardation, accounting for one third of all persons with a severe handicap. It is the best known but often one of the least understood of all mental handicapping conditions. The overall incidence has been estimated at 1/650 to 1/1000 births and mental handicap is typically further complicated by additional health problems. Many individuals with Down syndrome also have congenital heart disease, defects in vision and hearing, and suffer respiratory diseases and leukemia (Wishart, 1988). As late as the 1950's, only 50% of babies born with Down syndrome survived through infancy, but subsequent advances in medicine have increased life expectancy dramatically. Unfortunately, with a longer lifespan come other problems that research is now just beginning to identify.

Background to the Problem

Individuals with Down syndrome often display an array of physical features typical to the condition. Epicanthal folds, oblique palpebral fissures, broad bridge of the nose, protruding tongue, open mouth, square ears, muscular hypotonia, congenital heart disease, and varying degrees of mental retardation have been reported frequently as the

clinical manifestations of the condition (Grossman, 1983). Down syndrome (DS) is caused by a chromosome disorder in which an extra 21st chromosome, or in some persons, extra genetic material from chromosome 21 is present. Individuals with DS do not have defective genes, they simply have too many, and the presence of additional normal genetic material appears to affect normal physical and mental development.

Recently, research has linked Alzheimer's disease with its associated dementia to DS. Alzheimer's disease (AD) presents clinically with a progressive loss of memory and a deterioration in other specific cognitive functions, such as impairment in the ability to use words (aphasia), impairment in the ability to execute complex coordinated movements (apraxia), and impairment in the ability to recognize the significance of that seen, heard, or felt with the senses (agnosia). These specific deficits result in an overall decrease in intellectual functioning (Jolles, 1986). Alzheimer's disease is the leading cause of dementia in the elderly. It is estimated that at least 10 to 15% of the general population will develop this condition if they live past age 65, and AD is not a normal consequence of aging but a recognized pathological condition.

One current hypothesis suggests that a pathological segment on chromosome 21 is responsible for neuropathological changes in the brain of Alzheimer's patients. The presence of extra genetic material from

chromosome 21 found in the DS population, may increase the likelihood of Alzheimer-like changes. This leads one to question whether Alzheimer's disease will be an inevitable consequence of aging in DS.

One of the interesting aspects of DS is the premature appearance of a variety of age-related changes. Several studies confirm the presence of some aspects of accelerated aging beyond the third decade (Bauer & Shea, 1986; Eisner, 1983; Heston, 1984; Hewitt, Carter, & Jancar, 1985). In addition, the association between DS and AD that is so strongly indicated in the literature, suggests that the nature of age-related cognitive decline experienced by persons with DS, may be similar if not identical to dementia of the Alzheimer type (DAT) (Thase, 1988).

Neuropathological studies have contributed to the "hidden" identification of DAT. Diffuse cortical atrophy and neuronal loss, neuritic plaques in gray matter, extracellular and perivascular amyloid deposits, and neurofibrillary tangles have been reported as the key neuropathological features of AD (Thase, 1988). Evidence is now available that the gene for Beta amyloid protein which is responsible for amyloid deposits, plaques, and likely neurofibrillary tangles, is located on chromosome 21, thus implicating the pathological segment of chromosome 21 in both DS and AD (St Clair, 1987).

In studies of persons with DS 35 years or older, neuropathological changes of AD have been consistently documented (Bauer & Shea, 1986). Virtually 100% of DS persons carry neuropathologic changes of AD. Interestingly however, premature appearance and unusual density of plaques and tangles are seen in DS. They first appear in the second decade in DS, but in the fourth decade (tangles) and fifth decade (plaques) in persons within the general population (Kemper, 1988). In the fourth decade (age 30-39) in DS brains, plaques comparable to that in normal brains greater than 75 years of age are found, whereas the number of tangles further exceeds that level. It is noteworthy that the prevalence of Alzheimer-type neuropathology in the non-DS mentally retarded group has been reported to be consistent with the non-retarded population (Barcikowska, Silverman, Zigman, Kozlowski, Kujawa, Rudelli, & Wisniewski, 1989).

Surprisingly, while 100% of DS adults carry neurological evidence of AD, reports of the incidence of clinically defined dementia vary widely in the DS population. Thase (1988) reported that only 30% display DAT, while Dalton and Crapper-McLachlan (1986) found dementia in about 85% of the cases of DS showing the neuropathology of AD. Although reports of the incidence of DAT fluctuate greatly, the question remains as to why the neuropathology of AD does not always translate to overt

behavioral symptoms within this group. Thase (1988) suggested that there appeared to be an uncoupling of brain pathology and dementia in DS.

In the general population, DAT begins with learning and memory deficits and slowly progresses to IQ loss, EEG changes, and seizures (Dalton & Crapper-McLachlan, 1986). Toward the terminal stage, increased muscle tone, defective upward gaze, abnormal sucking and grasping reflexes, incontinence, and the loss of useful motor activity are common. In comparison, the most frequently reported clinical-functional expression among DS cases are the appearance of seizures, personality changes, apathy and inactivity, loss of conversational skills, incontinence, and EEG abnormalities. Memory loss, depression and disorientation which are among the earliest signs of dementia in the general population with a diagnosis of DAT are among the least frequently reported clinical features in DS. Dalton and Crapper-McLachlan (1986) reported "that no single clinical feature or functional change has been universally reported for dementia in DS with confirmed histopathology of AD at postmortem" (p. 664). Cognitive functions such as memory are perhaps too subtle to be noted reliably with this group. Rather, global changes in personality, self-help skills, and activity level are most commonly reported as typical functional changes.

Nature of the Study

Research into the association between DS and AD has largely focused on neuropathological changes resulting from the disease process. While postmortem confirmation of AD is important, it comes too late to identify and serve those who struggle with the disease. Screening devices are available to assess AD in the general population but appear to be inappropriate for detecting change within the mentally handicapped population. Behavioral indicators showing decline in functional skills, are more likely to aid in the diagnosis of AD with this group. Therefore, research that more clearly defines the aging process within the DS population is needed to determine if changes in adaptive functioning do occur increasingly with age, and if so, to define when within the lifespan changes become apparent.

A preliminary study of 143 adults with DS residing in Michener Centre, an institution serving 1200 mentally handicapped adults, compared pre- and post measures of cognitive functioning and adaptive behavior from existing psychological assessment records (Ho, 1989b). Consistent use of a single test instrument, as well as comparison of DS adults with mentally handicapped non-DS persons would provide more conclusive results of age-related decline in these two groups. As institutionalized populations tend to be of a lower level of intellectual and adaptive functioning, measures of intellectual functioning are less

useful to determine cognitive decline within this group. It is possible that many residents would slip below the testable range on available intellectual assessment instruments. Measures of adaptive functioning are likely more appropriate indicators of change within this population. As a paucity of research exists in the area, this study investigated age-related changes in adaptive functioning in institutionalized adults with DS as compared to their non-DS mentally handicapped agemates.

Research Question

Do adaptive skills in Down syndrome adults show a greater decline with age than adaptive skills of non-Down syndrome mentally handicapped adults?

CHAPTER II

Review of the Literature

Research detailing the impact of AD on DS stems from two major disciplines. Medical literature investigating the disease process at a molecular level has contributed by far the greatest volume of research. Advances in knowledge on this front promise to more clearly define the impact of genetics on the initiation and course of the disease. Findings presented in the psychological literature, although far less in volume, outline the incidence and outcomes of the disease on the total person. Contradictory findings between those present on a molecular level, and those observed at a behavioral level have fueled the controversy which surrounds the link between DS and AD.

In the following literature review, research over the past decade which pertained to adaptive functioning of persons with DS is presented. The chapter is divided into four sections: (a) aging in DS, (b) Alzheimer's disease, (c) link between DS and AD, and (d) assessment of adaptive behavior.

Aging in Down Syndrome

Much has been written about the etiology and outcomes of DS. Berini and Kahn (1987) synthesized what is known about the causes of DS. They reported that the excess of

21st chromosomal material, which typifies DS may result from any of several mechanisms. Trisomy 21 with its 47 chromosomes occurs as a result of a cell division error called non-disjunction, when during division and duplication of ova or sperm cells a pair of chromosomes fails to separate, leaving both number 21 chromosomes in one cell. Fertilization adds an additional 21st chromosome to the embryo, bringing the total to three instead of the normal two. As women age they are at increased risk for errors during this meiotic process and therefore for offspring with this type of DS.

Translocation DS with its 46 chromosomes is caused by a rearrangement of chromosomal material. This results from breakage of a number 21 chromosome and subsequent reattachment of the broken portion to another chromosome. In this case the total number of chromosomes is normal, but extra material from the 21st chromosome is present.

Mosaic trisomy with 47 chromosomes in some cells and 46 in others typically results from non-disjunction after fertilization, where an error in cell division in the developing embryo itself adds an extra chromosome to selected future cells. Mosaicism, however, may also occur as a result of an error in meiosis in the parent ova or sperm cell yielding trisomy 21 at conception with subsequent loss of a 21st chromosome during early division of the fertilized egg. The developing embryo then displays a

combination of cells containing either 47 or 46 chromosomes. Mosaicism, a rare form of DS, results in individual outcomes less predictable than those associated with a trisomy 21 or translocation etiology.

Typical clinical features result from an increase in genetic material from the 21st chromosome. Several studies have identified the most common signs of the syndrome. Pueschel (1988) reported small skull, eyes, nose, and ears, narrow ear canals, small oral cavity with enlarged tongue and adenoids, a short neck, short extremities, and occasional absence of the 12th rib to be characteristic of the condition. More specifically he detailed prominent epicanthal folds and oblique palpebral fissures as a common finding in 97% of youngsters with DS. Abnormal structures around the mid-facial region contribute to typical characteristics. Small oral cavity and enlarged tongue, in conjunction with narrow nostrils often result in mouth breathing and an open-mouth, droopy chin appearance.

Cunningham (1982) indicated that many unusual physical and behavioral characteristics of DS are related to differences in the skeletal system. At birth the arms and legs of the DS infant are usually within the normal range. However, slow growth of the skeleton produces noticeable retardation in height by 4 to 6 years of age. Additional symptoms are seen in a smaller pelvis with less developed bones, and flatter, wider iliac wings than found in the

general population. Poor muscle tone and flexibility in the tendons contribute to frequent gait abnormalities and often to flat feet in DS persons. Curvature of the fifth finger and a larger than normal space between the first and second toes have also been recorded in 96% of those with DS. In many DS persons, a simian line, or single crease across the palm, is found and has attracted attention as a diagnostic sign in DS (Pueschel, 1988).

Medical problems commonly associated with the syndrome have been reported in the literature. Pueschel (1987) reviewed health problems typical to infancy, childhood, and adulthood. He reported that an increased number of congenital abnormalities are found in DS. Gastrointestinal malformations are demonstrated in tracheoesophageal fistulas, and an imperforate anus more often in DS infants than in the general population. Congenital heart disease has been observed in 30 to 45% of children with DS and is the major contributor to the higher infant mortality rate seen in DS children in the first two years of life. The two most common cardiac abnormalities are presence of an atrioventricular canal, or a ventricular septal defect. Approximately 30% of those with heart defects have been reported to have multiple abnormalities (Odell, 1988).

Vision problems, including congenital cataracts, strabismus, nystagmus, refractive errors, and retinal problems have been commonly reported in DS individuals.

Niva (1988) found that cataracts were observed in more than 50% of patients with DS. In addition, hearing impairments are frequently reported. Previous investigations reported hearing impairment in 15 to 50% of the DS group. However, new assessment techniques have resulted in updated estimates that indicate 78% of individuals with DS display some hearing loss (Odell, 1988; Pueschel, 1987). Of this the vast majority of individuals suffer a conductive hearing impairment with the degree of hearing loss reported to be mild to moderate. Persistent fluid in the middle ear, a result of narrow ear canals and improper drainage was cited by Odell (1988) as a primary cause of hearing impairment.

Orthopedic problems reported in DS persons are attributed to generally low muscle tone rather than to congenital abnormalities (Odell, 1988). Cervical spine instability, likely related to ligament laxity and abnormalities of the vertebrae themselves are found in 10 to 20% of individuals with DS.

There are numerous reports in the literature of thyroid dysfunction in DS (Cunningham, 1982; Odell, 1988; Pueschel, 1987). The prevalence of thyroid problems are reported to vary from 3 to 50% depending on the age group examined. In older individuals with DS as many as 50% experience thyroid abnormalities, usually hypothyroidism (Pueschel, 1987). Symptoms of hypothyroidism include delayed growth and short stature, obesity, lethargy, and dry skin which because of

their frequency encourage us to view these symptoms as typical features of DS. As a result, a diagnosis of hypothyroidism could be easily overlooked. Finally, seizure activity has been reported as frequently in youngsters with DS as in the general population. Beyond age 20, however, incidence of grand mal seizures escalates among the DS population (Odell, 1988). Thus, the symptomatology and health problems which accompany DS have been frequently detailed.

Investigations into the unique developmental process present in DS also have been recorded in the literature. From birth, the rate of development appears altered in DS. Kemper (1988) consolidated findings in the field of neuropathology with regard to aging in DS. He reported that at birth although brain weight is approximately normal, by two years of age a clear decrease in expected brain weight is noted. Likewise, a slight decrease in head circumference at birth translates to a decline that is three standard deviations below expectation by two years of age. Further, a decrease in the size of the cerebellum and brain stem, a deficit in small localized neurons, and a variable pattern in large neurons are found in DS. Myelination, conversely, is reported to be on schedule at birth and during early childhood. Kemper stated that:

> It appears that the weight of the brain, skull size, myelinization, and development of individual cortical neurons are within the normal range up

> until birth. Then there is a progressive curtailment of brain development during the first postnatal years. This curtailment is evident in brain weight, head circumference, and development of individual neurons. (1988, p. 276)

This pattern of development, a curtailment of growth and maturation of the brain in infancy and early childhood, has also been noted in other aspects of postnatal growth and maturation. Kemper reported a statistically significant decrease in body weight and length, in IQ, and in scores on the Gesell Development Scales. At birth, body weight and length in DS were found to be 0.5 standard deviations lower than control neonates, and by 7 years of age 1.5 to 2.0 standard deviations less than non-DS children. The reason for progressive curtailment of growth and maturation of both brain and body tissue in infancy and early childhood is unclear. Kemper (1988) indicated that it resembled that seen in metabolic diseases like phenylketonuria where the placenta compensates for the deficit during pregnancy, which then becomes more apparent following birth.

Wishart (1988) emphasized the importance that expectations play in the ultimate development of an individual. She found that not only the general population, but also parents of DS infants, believed that their developmental potential was severely limited. Given poor muscle tone which restricts the infant's ability to explore and to initiate learning situations, in combination with low

parental expectations and a learning style that doesn't maximize learning, it becomes obvious why a decline in the rate of development is evident from birth.

Wishart (1988) reported earlier findings from a 1979 Edinburgh Longitudinal Study that attempted to provide a detailed description of the natural course of early learning in children with DS. Results indicated that qualitative differences in learning patterns reflected an approach to learning which was distinct from that seen in normal development. The DS children failed to consolidate recently acquired knowledge and were further disadvantaged by being less able to build on newly acquired information. Wishart found that social behaviors to engage the experimenter were a mechanism by which the DS person "switched out" of the learning task for social reinforcement and avoidance purposes. She reported that:

> The results from the Edinburgh Study suggest that while learning ability in DS is not as severely impaired as previously thought, poor consolidation of newly acquired understanding plus a tendency to under use the ability that is present, combine to limit achievement of developmental potential in DS. (1988, p. 42)

The increasing gap in DS and non-DS development may be due to deficiencies in learning, and manipulation of motivational factors in DS during development may be the key to enhancing learning and development. Gibson (1966) delineated growth curves of general intellectual

development in DS to support the position that their development is best characterized by discrete periods of steady development interspersed with plateaus during which little intellectual development occurs. Evidence such as this lends support to the idea that developmental processes in DS persons are not only delayed but also different qualitatively from non-DS individuals.

Although several studies report slower development in the early years, investigations into development in DS adults claim accelerated aging in the later years. For more than a century it has been thought that persons with DS age more rapidly than those who do not have this chromosomal disorder. Physical evidence supporting this claim is found in cataract formation, skin and adipose tissue changes, loss and greying of hair, early menopause, testicular atrophy, and the presence of cells seen post-menopausally (Gibson, 1978).

Sinex (1986) reported that the death rate at 60 years of age in DS corresponded to the death rate in the normal population at age 81, while Thase (1982) noted that mortality curves for DS between 40 to 60 years were similar to those for the general population aged 60 to 80 years. As well, a high proportion of conditions associated with old age were found in DS persons beyond their late 30's (Cunningham, 1982). St Clair and Blackwood (1985) in a study of P300 evoked potentials in persons with DS and in

non-handicapped controls reported that the pattern of change with increasing age was identical across groups. In DS persons however, the pattern was brought forward by many years indicating that the aging process was not more rapid, accelerated aging - but simply began earlier, premature aging.

Recently, reports in the literature have focused on the high prevalence of AD in adults with DS. Using a premature aging hypothesis, AD could be viewed as the inevitable result of early aging in DS. Although studies have found that the risk for AD increases with age to the extent that 25% of the general population over age 80 display the disease, it is unclear as to whether DS persons affected by AD have aged earlier than those unaffected. Therefore, it is more likely that in DS a specific disease process leads to AD, rather than a premature aging process.

Alzheimer's Disease

Alzheimer's disease is a progressive degenerative brain disease first identified by Dr. Alois Alzheimer in 1906. In the general population, more than 4% of persons aged 65 or over display senile dementia, of whom 50 to 65% have AD (Berini & Kahn, 1987). Khan (1986) indicated that the prevalence of the disease increased with age - 2.3% of persons 65 to 70 years old, 3.9% of 70 to 80 year olds, and 22% of those beyond 80 years display AD.

Reports of the rate of progression of the disease vary in the literature. Khan (1986) claimed death occurred within 6 to 12 years after onset. Ninos and Makohon (1985) cited a range of a few months to 20 years with a mean of 7 years diagnosis to death. Koss (1988) reported that different rates of disease progression and symptom severity may be related to age at onset. Patients affected before age 65 have faster disease progression and more widespread abnormalities (e.g., more frequent aphasia, apraxia, and agnosia) than patients affected after 65 years of age. Wright and Whalley (1984) also claimed that early onset was associated with greater genetic risk and severity of abnormality. Ortof and Crystal (1989) reported the rate of progression of AD to be variable and often unpredictable. However, they found that adults younger than age 65 at onset of illness did not progress significantly faster than did subjects with age of onset greater than 65 years. They concluded that age of onset, duration of illness, and family history of dementia have no significant influence on rate of progression. Similar findings were reported by Becker, Huff, Nebes, Holland, and Boller (1988).

The symptoms of AD are primarily behavioral and criteria used to diagnose the disease have been drawn from the American Psychiatric Association Diagnostic and Statistical Manual III (DSM III). According to the DSM III, a diagnosis of primary degenerative dementia can be made

when a loss of intellectual abilities, memory impairment, and at least one of the following are present: impairment of abstract thinking, impaired judgement, disturbances of higher cortical function (i.e., aphasia, apraxia, or agnosia), or personality changes.

Reisberg (1984) catalogued the stages of cognitive decline into 7 levels of cognitive function marked by recognizable clinical characteristics. The levels progress from: (1) normal - no cognitive decline with age; (2) normal aged forgetfulness - slight memory impairments associated with normal aging; (3) early confusion - decline in cognitive abilities begin to interfere with work and in social settings; (4) late confusion - symptoms of early AD make ability to handle routine marketing and financial tasks difficult; (5) early dementia - memory lapses, difficulty choosing clothing, anger and withdrawal; (6) middle dementia - fear of bathing, lack of memory about toileting and required assistance with both, agitation and paranoia; and (7) late dementia - inability to speak, walk, or chew. His contribution therefore, was to catalogue the progression of cognitive decline from normal, unaffected aging through the severe stages of AD.

Investigators have described the signs and symptoms of AD, however discrepancies exist between the progression of symptoms seen in the general population and those seen in the DS population. It is noteworthy that behaviors which

signify the possible onset of AD and cause problems for those who associate with Alzheimer's victims differ between these two groups. Following Reisberg's sequence, confusion in routine financial tasks highlights the problem in the general population, whereas lack of memory about toileting and bathing activities signify possible disease onset in DS. This underscores the idea that cohorts and caregivers may be sensitive to different aspects of daily living for different groups.

Newroth and Newroth (1980) charted 3 stages of decline in DS persons. Stage one was characterized by impairment of memory including diminished abilities in attention span, decision making, and judgement. Time disorientation, spatial disorientation with loss of the sense of direction, and inability to place the body in a correct position for an activity were reported. Lack of spontaneous movement and balance, diminished fine motor control and coordination, and diminished communication skills were also noted. When translated to daily living skills, the DS person with early AD demonstrated poor memory for names or objects, loss of interest in personal hygiene and appearance, and inappropriate dress for social functions. Loss of sensory feedback as to time of day, weather, and appetite, as well as difficulty in focusing and grasping were observed, such that at mealtime food spills resulted from trouble in finding, placing food on the fork, and locating the mouth.

Decreased body movement, alertness, balance, and eye contact, and an inability to button, dress, cut meat, and self-feed were noted. Finally, the loss of speech, hearing, significance of verbal communication, and reading and writing skills were reported. Symptoms in stage two according to Newroth and Newroth resulted from physical deterioration of the brain via increased size of ventricles, and brain shrinkage. Perceptual failure in which the individual lost the ability to recognize the significance of that seen, heard, or felt, as well as progressive speech and language difficulties were common. Movement disorders that interfere with mobility and self-feeding, the beginning of seizure activity, muscular trembling, increased muscle tone, and incontinence were noted. Emotional lability with frequent fear responses were also characteristic. Stage three was represented by complete dementia, with seizures, incoherent muttering, and poor response to people or other stimuli. Death usually occurred as a result of secondary infection caused by lack of movement. Personality changes were reported to follow the progression of the disease as early frustration and irritability translated to fear, expressions of sadness then euphoria, and finally despondency and withdrawal from others (Newroth & Newroth, 1980).

Several approaches to assessment of DAT have been suggested in the literature. Kertesz, Appell, and Fisman

(1986) suggested that accurate assessment of language was one of the most reliable predictors of the stages of AD offering insight into cerebral organization of language, semantic access, and memory involvement throughout disease progression. They reported that memory and cognitive decline were more significant initially than language impairment, but that aphasia was the presenting feature in many AD patients. Reading and writing were relatively more impaired than oral language they found. Tariot, Sunderland, Murphy, Cohen, Weingartner, and Makohon (1985) presented a theoretical perspective on how memory fails in AD. They suggested that both episodic memory and semantic memory function declined in AD in contrast to other dementing conditions.

Doyle, Dunn, Thadani, and Lenihan (1986) described the results of a functional assessment on hospitalized AD patients. From a review of self-care scores they found the greatest independence in function were in communication, ambulation, and transfer - functions which were maintained despite a severe level of dementia. The most severely impaired functions were daily living activities requiring complex movements such as grooming, dressing, hygiene, and eating. They also remarked that although fluency of speech was often retained, comprehension and ability to read and write were often lost or severely impaired. Doyle and associates concluded that in AD, the most impaired functions

were those that required fine motor skills, memory, and recognition; whereas skills which required gross motor activity remained intact initially despite severe mental deterioration. In a similar review of assessment of functional status with AD patients, Winogrond and Fisk (1983) attempted to determine to what extent cognitive function in DAT was associated with behavioral function. They concluded that a strong statistically significant association between cognitive and behavioral functions, and between morale and behavioral functions existed.

Cummings and Benson (1986), to distinguish DAT from other dementias, presented an inventory of diagnostic clinical features that include aphasia, amnesia, abnormal cognition and visuospatial skills, personality alterations, and motor system abnormalities. They reported that motor function remained normal until the final stage of the disease. Kaye (1988), however, targeted disorders of the motor system as markers of the anatomical and neurochemical changes occurring throughout progression of the disease. He indicated that knowledge of underlying physiological decline would be valuable in planning treatment. Many studies have outlined the clinical course of the disease.

The etiology of AD is not known but several causal theories have been proposed. Aluminum toxicity, viral infection, imbalance of neurotransmitters, enzyme deficiency, and vascular incompetence have been reviewed by

Glenner (1985) as possible contributors to the condition. Evidence for genetic contribution was confirmed in 1987 when the gene for Beta amyloid protein which is responsible for amyloid plaques, amyloid deposits, and possibly neurofibrillary tangles was located on chromosome 21 (St Clair, 1987). The link between AD and DS became more understandable as a result of this finding.

Research has found that AD is expressed not only on the behavioral level, but also on anatomical and biochemical levels (Schweber, 1985). Structural abnormalities in the brain of AD patients include neuronal plaques, located outside nerve cells and consisting of a core of amyloid surrounded by debris from degenerating neurons, and neurofibrillary tangles or twisted fibres within neurons or at synaptic terminals. The distribution of plaques and tangles has been found to correspond to cell losses and to correlate with the degree of dementia reported. Events that follow increasing losses of cortical neurons and accumulations of amyloid include impaired synthesis of neurotransmitters such as acetylcholine, reduced cortical blood flow, and reduced uptake of glucose in the parietal, temporal, and hippocampal areas of the brain. Cognitive decline and behavioral manifestations of AD follow as eventual outcomes of the genetic trigger. Although research has largely dispelled the idea that dementia is the product of normal aging, Sinex and Myers (1982) claimed that in

familial cases, AD was an inherited disease whose penetrance was a function of aging.

Link Between Down Syndrome and Alzheimer's Disease

An association between DS and dementia has long been known. Jervis (1948) described the clinical deterioration associated with Alzheimer-like changes at post-mortem in a number of people with DS. He stated that remarkable changes in behavior occurred in cases of "mongoloid idiocy" in the fourth or fifth decade of life. Loss of interest in daily tasks, a striking change from cheerfulness to sullenness, loss of personal habits of cleanliness, and increasing behavior problems preceded tremors and gait disturbances. Jervis concluded that with the exception of the age of onset, the clinical and pathological manifestations were those of senile dementia.

Following these early reports, most relevant research focused on establishing similarities between neuropathological changes in the brains of aging DS persons and the plaques, tangles, and degeneration characteristic of AD. By the early 1970's, the link between these two disorders was clearly established and on the basis of post-mortem data it was argued that all people with DS over the age of 35 had the neuropathological features of AD (Dalton, Crapper, & Schlotterer, 1974; Heston, 1984; Miniszek, 1983; Wisniewski, Dalton, Crapper-McLachlan, Wen, & Wisniewski, 1985). With the advent of antibiotics, a dramatic increase

in average life expectancy in DS persons occurred (Newroth & Newroth, 1980). Thase (1982) has shown that from a life expectancy of approximately 9 years in 1929, a mean life expectancy greater than 30 years is now reported with 25% of the DS population living to the age of 50. This increased longevity has given rise to a population of elderly persons with DS not previously seen.

Neuropathological studies have provided the most consistent and convincing evidence of Alzheimer-like changes in the brains of DS persons. Since senile plaques and neurofibrillary tangles are present in normal older people and tend to become more numerous with age, early investigators attributed their occurrence in DS to the premature aging phenomenon. Several studies dispute this claim however. Wisniewski, Wisniewski, and Wen (1985) stated that it was not only the presence of large numbers of plaques and tangles, but their distribution within the brain that contributed to dementia. Although they were unclear as to why plaques and tangles develop 20 to 30 years earlier in persons with DS, they suggested that perhaps all pathological changes, including decreased brain weight, brain atrophy, interference in synaptic transmission, and related biochemical changes, combine to bring about the early expression of DAT in DS. They concluded that a correlation existed between dementia, density of plaques and

tangles, and age, and that dementia is present three times more frequently in DS than in the non-DS population.

In a follow-up study, Wisniewski and Rabe (1986) reiterated that contrary to speculation, they found no evidence that the presence of a few plaques and tangles were associated with mild age-related decline in intellectual function. Their data indicated, rather, that a threshold number of plaques and tangles must be exceeded before the appearance of dementia and that DS individuals appeared to have a higher threshold for dementia as measured by plaque and tangle counts.

Heston (1984) also found that although neuritic plaques and neurofibrillary tangles were most prominently seen in the hippocampus and cerebral cortex, to be significant diagnostically they had to be present in substantial numbers. Findings presented by Rafalowska, Barcikowska, Wen, and Wisniewski (1988) indicated that plaque numbers in both temporal and occipital lobes of AD and DS persons were significantly higher than that of normal old people but there was no difference between AD and DS. Similarly Mann (1988) stated that pathological changes of DS at middle age were qualitatively the same as those of AD at that age but that quantitative differences did occur. He concluded that "in pathological terms patients with DS at middle age do indeed have AD" (Mann, 1988, p. 99).

Neuropathological and clinical evidence clearly point to a link between DS and AD and prompted comment by Bauer and Shea (1986, p.144) that "In Down's Syndrome the reward for survival beyond age 40 is presenile dementia". Whereas a high proportion of those with DS develop neuropathological changes of AD, only a proportion develop definite signs of deterioration and display the clinical features characteristic of the later stages of AD. Dementia may develop in persons with DS as young as age 35 (Jervis, 1948) while others show no evidence of decline by the age of 59 years (Thase, 1982). In a prospective study, Dalton and Crapper (1984) found that 24% of DS persons over 40 years of age showed evidence of memory deterioration with an onset ranging from 42 to 60 years. No memory deterioration was found among DS subjects under 40 years, nor in mentally retarded control subjects. Lai and Williams (1989) in a more recent prospective study, found the prevalence of dementia in an institutionalized DS population to be 8% in persons 35 to 49 years, 55% in those 50 to 59 years, and 75% in those over 60 years of age. This follows the trend of increasing incidence of dementia with age reported in the general population but is as Wisniewski and colleagues (1985) indicated, three times the frequency, and 20 years premature.

Research has shown that the course of AD in the DS population varies from that found in the general population

(Dalton et al, 1974; Miniszek, 1983; Oliver & Holland, 1986; Wisniewski et al, 1985; Wisniewski & Rabe, 1986). It is noteworthy that the majority of these studies focused attention on cognitive changes typical to AD. In 1974 Dalton and Crapper (1984) stated that clinical features of the first two stages of the disease were rarely reported in persons with DS. Their study compared groups of institutionalized "young Down's" (19 to 23 years), "Intermediate Down's" (39 to 43 years), and "Old Down's" (44 to 58 years) with non-DS mentally retarded subjects matched for age on a visual retention task using a Skinnerian paradigm. They found evidence of memory loss in the group of "old" DS persons that was similar to that exhibited by non-retarded people with AD. Absence of a decline in function in the non-DS groups indicated that the results were unlikely to be simply age-related. Dalton and colleagues concluded that poor retention test performance was an indicator of early AD, however this memory defect was normally masked in aging DS persons due to profound language, intellectual, and behavioral limitations. Although they make a valid point, this study may be criticized from the perspective that they assume that a visual retention deficit truly represents cognitive decline in a population known to suffer increasing visual impairments with age.

Thase, Smeltzer, and Maloon (1982) surveyed clinical signs and symptoms of dementia in an institutional population with DS and in control residents matched for age, sex, IQ, and length of institutionalization. From 170 DS residents, a random sample of 10 subjects per decade between the ages of 25 to 64 were selected and evaluated via neurological examination, neuropsychiatric interview, and subtests measuring cognitive functioning. They reported that overall 75% of DS subjects had one or more symptoms of dementia compared to only 30% of control subjects, while 45% of DS but only 5% of non-DS persons displayed the full syndrome of dementia. The cross-sectional design of this study was recognized as a major limitation. A longitudinal design to follow the incidence and course of dementia over time would have produced more useful results.

Assessment via the Stanford-Binet Intelligence Scale regularly administered to institutionalized DS residents indicated that significant associations were found between intellectual deterioration and decreased visual acuity, and hearing loss - typical features of aging in DS adults (Hewitt, Carter, & Jancar, 1985). A high level of behavioral functioning and few behavioral problems that could not be attributed to prolonged institutionalization were found. In question here is whether the DS group differed from their non-DS cohorts and whether the Stanford-Binet, a verbally loaded assessment instrument, can

adequately assess dementia in the handicapped population. Miniszek (1983) claimed that the structured lifestyle typical to institutional living did not place intellectual demands on residents to meet their daily needs. Early signs of AD were thus easily masked. Reasons for the reported absence of behavioral deterioration in performance of self-care skills were unclear however.

Problems associated with the assessment of cognitive decline in generally low functioning institutional populations has resulted in an altered research focus. In a study by Lai and Williams (1989), dementia was judged to be present when a functional decline occurred in areas such as orientation, memory, verbal and motor skills, and self-care abilities. Institutional and community-based DS persons over 35 years of age were evaluated over an 8 year period by a standard neurological assessment, caregiver reports and records of level of functioning, and behavioral observation.

Three phases of clinical deterioration in DS were recognized. Initially memory impairment, temporal disorientation, and reduced verbal output were found in higher functioning persons with DS while apathy, inattention, and decreased social interactions were the first indicators of dementia in more retarded subjects. Both groups were reported to get lost in and around the residence. The second phase was typified by loss of self-help skills, such as dressing, toileting, and using food

utensils, slowed shuffling gait, a decline in workshop productivity, and seizures. Finally, non-ambulation, flexed postures, incontinence, and pathological sucking and grasping reflexes were characteristic of phase 3.

The significant contribution of this study was to correlate the first signs of dementia in DS with the later stages of DAT in the general population, and to identify a higher incidence of seizures in DS dementia than in the general population with DAT. Lai and Williams stated that even in those with mild to moderate retardation, poor language skills and memory disturbances were commonly masked. Rather, the first symptom was a change in personality such as irritability. In the severely to profoundly retarded group, need for more assistance in toileting, dressing, and eating was often the first clue to their decline. They concluded that any degree of retardation makes detection of early DAT difficult due to the neuropathological features of AD being superimposed on an already developmentally abnormal brain. This offers an explanation for the seemingly altered course of AD in persons with DS.

Research has adequately demonstrated the link between DS and AD. Nevertheless, the overwhelming neuropathological evidence in all DS persons over age 35 does not demonstrate 100% compliance with reports of the clinical expression of AD. Whether the course of the disease is truly different in

DS persons, or just masked by already altered brain development is unknown. Research to assess cognitive decline in DS has followed creative avenues but has not offered the answers expected. Perhaps a shift in focus toward areas of loss of functional skills would help to clarify the presence and pattern of AD expression in DS.

Assessment of Adaptive Behavior

Since the founding of the American Association on Mental Retardation (AAMR), its members have directed their energy to distinguishing mental retardation from other handicapping conditions. Previously, classification of mental retardation was solely based on performance on measures of intellectual function. In 1961, the AAMR formally added an adaptive behavior component to the measurement of intelligence in the definition of mental retardation. The current and most widely used definition states mental retardation as: "significantly subaverage general intellectual functioning existing concurrently with deficits in adaptive behavior, and manifested during the developmental period" (Grossman, 1983, p.1).

This significant move on the part of the AAMR has resulted in increasing emphasis to not only define adaptive behavior but to formally measure it through standardized procedures. Whereas the measurement of intelligence attempts to objectively assess potential for academic achievement through verbal and quantitative learning,

abstract thinking, and ability to generalize, adaptive behavior emphasizes social competency through the current ability to cope with environmental demands which are non-academic in nature. Adaptive behavior is defined as "the effectiveness or degree with which individuals meet the standards of personal independence and social responsibility expected for age and cultural group" (Grossman, 1983, p.1). It is, therefore, a socially defined concept, the expectations for which vary with age.

Standardized assessment instruments have flourished in an attempt to tap abilities necessary to cope with the environment. Although a diversity of behavior domains are covered on these instruments, Meyers, Nihira, and Zetlin (1979) described seven behavior domains commonly recognized in adaptive behavior scales. They are: (1) Self-Help Skills - attention to immediate personal needs; (2) Physical Development - perceptual motor development and coordination; (3) Communication Skills - social use of language; (4) Cognitive Functioning - development and functional use of reading, writing, number, and time concepts; (5) Domestic and Occupational Activities - productive use of time in domestic and vocational activities; (6) Self-Direction and Responsibility - self-initiation of purposive activities; and (7) Socialization - interaction and cooperation with others. Within domains, related subdomains serve to differentiate further the presence or absence of everyday

proficiencies in functional abilities. Grossman (1983) made the point that, due to contributing factors unique to each person which result in within-subject variation, subdomain scores are a more valuable measure than a composite score reflecting general level of adaptive behavior.

Adaptive behavior scales have typically been divided into norm-referenced assessments, designed to discover how well an individual performs in relation to others of their same category, and criterion-referenced assessments, designed to compare achievement to an absolute standard. Of importance in criterion-referenced assessment is the need for the sequence of items to be functionally related, since it is often used to reflect the outcomes of training (Hogg & Raynes, 1987). Finally, Raynes (1987) made the point that, although difficult to assess on adaptive behavior scales, information on the reliability, validity, and standardization process aids interpretation of results.

Recently, The Pyramid Scales: Criterion-referenced measures of adaptive behavior in severely handicapped persons (Cone, 1984) has been developed. Because the scale is relatively new, a computer search failed to yield previous citations. In addition, J. D. Cone (personal communication, August 17, 1990) stated that, "I still do not know of references to similar work that I could mention, although numerous places are accumulating large amounts of Pyramid Scale data". However, on annual administrations to

700 clients at the Greene Valley Developmental Center in Tennessee, F. Lambert (personal communication, August 29, 1990) evaluated the test-retest stability of the total score and subdomains and found all correlations to be statistically significant at the .01 level. Further validation research is needed with this instrument.

Studies focusing on the prevalence, detection, and course of AD in DS populations have held as their primary focus assessment of decline in cognitive abilities over time. Given the problems with teasing out cognitive changes in initially low functioning institutional populations, several investigators directed their efforts to changes in adaptive behavior over time as more observable and functionally relevant indicators of the disease process.

Although observable changes in adaptive functioning have been noted, several alternate explanations for this change have been proposed in the literature. Francis (1970) investigated the effects of age in "young" and "old" institutionalized DS individuals and although a narrowing of interest in surroundings was found, concluded that behavioral changes were due to institutionalization rather than to aging. Likewise Fine, Tangeman, and Woodward (1990) reported increases in both adaptive and maladaptive behavior with deinstitutionalization of older adults undifferentiated by etiology. Krauss and Seltzer (1986), conversely, did not

find differences between adult and elderly mentally retarded persons living in institutional and community settings.

Libb, Myers, Graham, and Bell (1983) searched for factors which would account for variability among individuals with DS 3 months to 22 years of age. They indicated that IQ and adaptive behavior scores were positively correlated with parental level of education since more highly educated parents sought better health care and developmental experiences for their children. Declines in adaptive functioning were attributed to children of older parents for whom societal treatment and parenting practices were of the "old school". These results, however, might also reflect the greater incidence of trisomy 21 in the older parent group, and of mosaicism and translocation DS in the younger parent group.

Silverstein, Ageno, Alleman, Derecho, and Gray (1985) found that institutionalized DS adults displayed greater social competence and less maladaptive behavior than matched non-DS peers. Favorable expectations of service providers in eliciting more positive and less negative behaviors from their clients was the presumed cause. Finally, in a study of non-DS persons 65 years or older Hewitt, Fenner, and Torpy (1986) cited evidence for ongoing intellectual development until late middle age followed by a significant decline in 18% of persons. Intellectual deterioration was associated with deficient self-care skills and poor

orientation, unrelated to previous functioning level or prolonged institutionalization.

Several studies have attempted to delineate the course of AD in the DS population through examination of adaptive functioning. Lott and Lai (1982) indicated that of DS adults 32 to 64 years old referred for mental deterioration over a 2 year period, 100% showed personality changes and loss of independent daily living skills. Deterioration took the form of apathetic withdrawal in the home and vocational setting, and loss of spontaneous social interest. In almost every instance, these were first associated with a lessened interest in daily hygiene and need for increased prompting by caretakers. They claimed the motivation for performing these tasks failed before the actual loss of motor control. Secondarily, seizures in 53% of persons, deterioration of gait in 75%, and incontinence in 40% signaled a characteristic dementia syndrome in DS. Initial symptoms of recent memory, arithmetic, and visuospatial abnormalities in the general population with AD, contrasted that found in DS. Although these skills would not likely lead to referral of DS persons for mental deterioration, Lott and Lai attributed this to assessment difficulties, and to differences in the topography of pathological changes in DS and non-DS brains.

Dalton and Crapper-McLachlan (1986) assimilated clinical reports of deterioration presented in other studies. They found seizures, personality changes,

incontinence, apathy and inactivity, loss of conversation, and loss of self-help skills cited in decreasing incidence. Memory loss and disorientation were least often reported among DS persons. They concluded that the clinical expression of dementia in DS could be attributed to AD, environmental factors, or an unrecognized illness confounded by overriding brain damage. Finally Fenner, Hewitt, and Torpy (1987), via a cross-sectional study using Stanford-Binet and Adaptive Behavior Scale (ABS) data, found that intellectual decline occurred in less than 1/3 of DS adults over age 35. Level of self-care skills remained reasonably high and behavior problems, such as inactivity, were infrequent and likely due to prolonged institutionalization. As a longitudinal comparison was not possible, this study reflects methodological problems associated with cross-sectional design. Cautious interpretation of the relationship between brain pathology and psychological function was therefore recommended.

Adaptive functioning has also been the vehicle through which investigators have attempted to reconcile the discrepancy between the neurological and clinical presence of DAT. Miniszek (1983) was among the few concerned with difficulties of early and proper diagnosis of dementia in low functioning DS persons. Through ABS assessment, he found that old DS persons were rated lower than their age-matched peers without DS. He further differentiated between

older regressed and non-regressed DS individuals by recording an extremely uniform pattern of abilities in the regressed group versus a greater range of abilities and diversity of profiles in those not regressed. He believed that changes in an individual's profile over the years might indicate AD during the first and second stages and recommended the ABS as a valuable diagnostic tool. Eisner (1983) similarly focused on the neurological/clinical discrepancy and concluded that although there was evidence for accelerated neurological aging, a slight relationship between the degree of neurological changes and behavioral signs of senile dementia existed.

Silverstein, Herbs, Nausta, and White (1986) examined the effects of age on adaptive behavior and discovered that age did not differentially affect adaptive behavior of mentally retarded persons with or without DS. Rather, reported declines in motor and eating areas and an increase in writing skills, which did not differ across etiology, were due to the initial low level of functioning of subjects (i.e., a floor effect), or to a lack of power in statistical tests used. Similarly Zigman, Schupf, Lubin, and Silverman (1987), in a study of 2,144 persons with DS and 4,172 without, found adaptive competence declined with increasing age to a greater extent in DS individuals than in their peers. Age-related deficits were clearly seen only in DS persons over 50 years of age but the pattern of age-related

decline was not affected by functioning level or residential placement. They indicated that the most sensitive manifestations of regression in this population were found in behavioral domains related to activities of daily living, rather than in the cognitive domain, and suggested DSM III criteria for diagnosis of dementia be modified to reflect the restricted cognitive repertoires of mentally handicapped persons.

Synthesis of past research into well founded statements detailing the presence, prevalence, age at onset, and clinical course of DAT in DS is yet premature. Contradictory research abounds, confounded by methodological limitations of popular cross-sectional designs, investigation of low functioning institutional populations, small sample sizes, inadequacies of cognitive assessment procedures in low functioning persons, and discrepant expectations for clinical expression based on neuropathological evidence. The present study attempted to sidestep these problems in part, proposing instead to examine age-related changes in adaptive function in a total population of DS adults and their matched peers via longitudinal as well as cross-sectional methodology. Availability of prior adaptive skill assessments necessitated reliance on an institutional population of mentally handicapped persons. Nevertheless, results of the study were expected to have some generalizability.

CHAPTER III

Method and Procedures

The following chapter will examine the technical aspects of this research study. Subjects and instrumentation will be described. This description will be followed by an outline of data collection procedures and data analysis techniques that were applied to answer the research question outlined previously.

Subjects

A total sample of 280 subjects (140 adults with Down syndrome, and 140 adults without Down syndrome) was obtained from mentally handicapped residents of The Michener Centre. Diagnosis of DS made previously by physicians was primarily based on the presence of physical features typical to the condition. In 24% of the cases (34 of 140 subjects), chromosomal studies had confirmed trisomy 21.

From a 1988 population of DS adults, those assessed in 1985-89 with The Pyramid Scales were selected. Although 145 adults with DS were originally identified as potential research subjects, 5 individuals had not been assessed with The Pyramid Scales. The final sample of 140 adults however, represented 97% of the target DS population.

Subjects with DS were assigned to one of two groups according to their age at initial assessment. One group was

less than 40 years of age (n = 98), the other group was 40 years of age or older (n = 42). Within each group subjects were further subdivided by functioning level at initial assessment into a mild/moderate (m/m) or severe/profound (s/p) classification of mental retardation. Breakdown of the DS group by functioning level (F/L) was reported as mild (n = 12), moderate (n = 38), severe (n = 38), and profound (n = 52).

Of the DS target group, 19 subjects carried a medical diagnosis of legal blindness ranging from 20/200 vision to total blindness. No subjects with a medical diagnosis of deafness, and no subjects with behavior problems severe enough to warrant transfer to a special Behavior Management residence were within the DS group. During the 1985-89 interval, 7 DS subjects initially assessed on the Pyramid Scales died or were discharged from the institution. Their scores were retained in the DS group. Of the DS group, 76 were male and 64 were female.

Comparison groups of non-DS adults with a 1985-89 measure on the Pyramid Scales were drawn from the general institutional population of approximately 1100 mentally handicapped residents. Residents who had died or been discharged prior to April 1990 were not included in the subject pool. Included in this population were individuals with cerebral palsy, epilepsy, a variety of genetic syndromes excluding DS, and other neurological impairments.

DS and non-DS subjects were matched (1:1) on the basis of age (birthyear), functioning level (mild, moderate, severe, profound mental retardation), gender, sensory handicap, and residential unit at initial assessment. Subject characteristics were ranked and matching proceeded systematically down the hierarchy until as close a match as possible on the stated parameters was found. All subjects were matched on functioning level. Mismatches occurred on 14 of 140 DS subjects on birthyear, however, in each case birthdates were within 6 months of each other in the succeeding or following birthyear. A mismatch on gender occurred in 6 of 140 subjects. The 19 legally blind DS subjects included in the study were matched subject to subject if possible ($\underline{n}$ = 5), or within subgroups when a partner also meeting all other parameters could not be found ($\underline{n}$ = 14). Prevalence of blind subjects per subgroup was as follows: (1) DS <40 m/m ($\underline{n}$ = 2); (2) DS <40 s/p ($\underline{n}$ = 9); (3) DS 40+ m/m ($\underline{n}$ = 0); (4) DS 40+ s/p ($\underline{n}$ = 8) with equivalent numbers in corresponding non-DS subgroups. Deaf residents were excluded from the non-DS group.

Although matching per residential unit at initial assessment was attempted, this parameter was seldom satisfied. During the 1985-89 target period, all residences within the Michener Centre were renamed. Several units underwent kitchen and bathroom renovations necessitating the closure of some, opening of others, and transfer of

clientele to available beds within the institution. Matching on this parameter was approximated, therefore, by pairing DS and non-DS subjects residing at initial assessment in comparable residential clusters. Any one of 23 group homes, college style dormitory residences with single or shared bedrooms, areas designated by medical needs, physical handicaps, or senior clientele, or residences providing congregate care in dormitory-dayroom facilities were combined on this parameter. Demographic characteristics of DS and non-DS subgroups are summarized in Table 1.

Instrumentation

Because this study was retrospective in nature, the instrument chosen for assessment of adaptive behavior was determined by the Michener Centre's existing psychological assessment program. A description of the instrument including methods of administration, scoring, reliability, and validity follows.

The Pyramid Scales: Criterion-Referenced Measures of Adaptive Behavior in Severely Handicapped Persons (Cone, 1984)

The Pyramid Scales is a criterion-referenced measure used to assess adaptive behavior in handicapped persons of all ages. It exists as a component in The Pyramid System, a total educational/training service delivery system for

Table 1 Demographic Characteristics of Down Syndrome and Non-Down Syndrome Subgroups

Subgroup	Years Institutionalized ± SD	Initial Assessment N	Initial Assessment Age* ± SD	Retest N	Retest Age ± SD
<40 m/m					
DS	20.7 ± 7.9	37	31.7 ± 4.6	4	37.0 ± 4.2
non-DS	15.2 ± 8.5	37	31.1 ± 4.7	15	35.3 ± 3.4
40+ m/m					
DS	28.4 ± 9.0	13	48.3 ± 3.9	6	51.0 ± 3.3
non-DS	28.7 ± 9.1	13	48.0 ± 4.0	8	51.6 ± 3.0
<40 s/p					
DS	19.6 ± 6.4	61	29.4 ± 5.0	31	32.4 ± 5.1
non-DS	19.7 ± 6.4	61	29.5 ± 4.9	30	31.9 ± 4.7
40+ s/p					
DS	26.1 ± 9.2	29	50.9 ± 7.9	15	53.2 ± 7.6
non-DS	28.4 ± 8.0	29	51.2 ± 7.6	11	53.9 ± 8.4

* Mean age in years

handicapped persons. In total The Pyramid System consists of a series of carefully interrelated components which include assessment, curricular, parent involvement, progress monitoring, and staff training materials. The Pyramid Scales assessment was used as a stand alone measure of adaptive behavior in this study.

Items in The Pyramid Scales are appropriate for very young or for very low functioning persons. It is useful for surveying adaptive skills in an institutional population serving a predominance of severely-profoundly developmentally disabled persons. Content covered by the scales is described by the 20 subdomains listed below:

I. Sensory Zone
   1. Tactile Responsiveness (TR)
   2. Auditory Responsiveness (AR)
   3. Visual Responsiveness (VR)

II. Primary Zone
   4. Gross Motor (GMOT)
   5. Eating (EAT)
   6. Fine Motor (FMOT)
   7. Toileting (TOIL)
   8. Dressing (DRES)
   9. Social Interaction (SOCI)
   10. Washing/Grooming (WASH)
   11. Receptive Language (RLAN)
   12. Expressive Language (ELAN)

III. Secondary Zone
   13. Recreation/Leisure (REC)
   14. Writing (WRIT)
   15. Domestic Behavior (DOME)
   16. Reading (READ)
   17. Vocational (VOC)
   18. Time (TIME)
   19. Numbers (NUM)
   20. Money (MON)

Each subdomain samples 8 behaviors ranked by level of difficulty. Each behavior consists of 4 component skills

also ordered from simple to complex. Individual items are scored on a 4 point scale which reflects increasing degrees of competence in performance of the particular skill. In addition, a subject may be scored as Physiologically Incapable, or as having No Opportunity to perform the item (Cone, 1984).

The Pyramid Scales may be administered in 35 to 40 minutes by any of several modes: interview mode, informant mode, or direct observation. Accuracy of data collected is dependent on either the observation or interview skills of the test administrator, and their familiarity with the subject and test instrument. Administration by persons familiar with the scales and who have been trained in their use is recommended. Responses are recorded on a standard answer sheet. Only subject responses directly observed by the informant or observer are credited.

Subdomain scores for all 20 scales are obtained by dividing the number of subitems scored by the number of subitems actually administered, excluding those rated as Physiologically Incapable, or No Opportunity to observe. The resultant ratio is multiplied by 100 to obtain a percentage competence score per subdomain.

Evidence for reliability and validity of The Pyramid Scales is available in the manual. Test-retest stability estimates for the same group of subjects on a mean retest interval of 11.85 months ranged from .58 for the Vocational

subdomain to .97 for the Fine Motor subdomain, with a mean of .88 for all scales and all age groups. The Kuder-Richardson 21 formula was used to compute internal consistency of each of the scores with a resultant coefficient mean of .92 across all scales (Cone, 1984).

Various types of validity have been established for the scales. The most extensively developed was content validity, ensured via review of over 250 behavior checklists with inclusion of various adaptive behavior categories from the most comprehensive of these (Cone, 1984). In addition, The Pyramid Scales follow guidelines suggested by the AAMR pertaining to the standardization of measures of adaptive behavior, and sampling of everyday proficiencies of self-care, communication, and social responsibilities (Grossman, 1983). Standard administrative and scoring procedures, an interpretive manual, and scores per subdomain or content area, rather than total score representing general level of social competence, were developed.

Construct validity was demonstrated in various groups of individuals across different settings. Scores were noted to be generally developmental with the scales arranged hierarchically (Cone, 1984). These data support the construct validity of this instrument. Acceptable measures of reliability ($r = .92$) and validity were also reported in data collected at The Michener Centre during the 1985-88 assessment cycle (Ho, 1989a, 1989b).

In the present study, measures of adaptive functioning were drawn from existing Pyramid Scale results recorded between 1985-89 and held in each subject's file. Assessments were conducted via interview with residential direct care staff, by members of Michener Centre's Psychological Services Department Assessment Team. Assessment team members were chartered psychologists, or psychological assistants all trained in administration of The Pyramid Scale and supervised by a specialist in psychological assessment. Administration of The Pyramid Scales, in conjunction with a measure of cognitive functioning, occurred routinely prior to each subject's birthdate in preparation for review of each Individual Program Plan. Under Michener Centre's internal assessment policy, psychological assessment of each resident is required every 3 years. The 1985-88 assessment cycle, therefore, resulted in Pyramid scale data recorded for most of the institution's residents. It was from this cycle that initial assessment data were drawn for all subjects. Determination of functioning level per subject was based on measures of intellectual and adaptive behavior functioning obtained at this assessment. Subjects were assigned to groups by etiology, age, and functioning level recorded at this time. Retest data were drawn from 1988-89 assessments.

Data Collection

Data collection proceeded in 3 phases. Phase one involved DS subjects with 1985-89 measures of adaptive behavior on The Pyramid Scales (total possible n = 145, final n = 140). The second phase involved matched non-DS subjects with 1985-89 measures on The Pyramid Scales (total possible n = 961, final n = 140). Only assessments conducted by the Psychological Services Assessment Team were accessed in phases one and two. Phase three involved all subjects with a possible 3 to 4 year retest interval assessed in 1989 by residential direct care staff (total possible n = 49, final n = 15 DS and n = 15 non-DS). Assessors had been formally trained in administration and scoring of The Pyramid Scales by the Psychological Services Assessment Team.

Phase One

A written proposal (see covering letter in Appendix A) outlining the pertinent details of this research study was submitted to the Research Sub-Committee of The Michener Centre Ethics Committee on March 1, 1990. Permission to conduct the study and to access data in the cumulative records of the residents was requested. Consent for ongoing assessment and programming given on admission to The Michener Centre, remained in place for the purposes of this study. Permission was granted by the Research Sub-Committee on March 30, 1990 subject to guidelines which included

ongoing consultation with the Psychological Services Assessment Team supervisor, and request for a progress report by October 31,1990 (see correspondence in Appendix A).

Data collection began April 2, 1990. Three computer printouts current to April 1, 1990 were drawn from Michener Centre's Central Client Records. One printout was an alphabetical listing of all Michener Centre residents (n = 1098) including birthdate, age, sex, functioning level, date of admission, significant medical condition (e.g., cardiac, epileptic, diabetic), and additional family or guardianship information. An identical printout comprised of only DS residents (n = 137) was also generated. Thirdly, a listing of all residents by year of birth, including name, current residential unit and functioning level was obtained. Three computer printouts were also drawn from Michener Centre's Medical Records. Listings of (1) all DS residents (n = 137), (2) all legally blind residents (n = 84), and (3) all deaf residents (n = 37) were generated by current residential unit.

Composition of the DS group as well as retrospective data was drawn from a 1985-90 spreadsheet of psychological assessment results compiled by the Psychological Services Assessment Team. The spreadsheet included, for each DS resident, full scale IQ on the Wechsler Adult Intelligence Scale - Revised (WAIS-R) or Peabody Picture Vocabulary Test

(PPVT), and total score plus subdomain scores on the Adaptive Behavior Scale (ABS) or The Pyramid Scales (PS).

All relevant data (e.g., demographic characteristics, test scores) were recorded onto a prepared data form (see Appendix B). Subjects were assigned ID numbers beginning with 001 and this number rather than their name was used on the data form to protect confidentiality and anonymity of information. Each DS subject was assigned to 1 of 4 subgroups based on age and functioning level at initial administration of The Pyramid Scales. For each subject a cumulative record of psychological assessment reports prepared by the Psychological Services Department was reviewed. At this time results obtained from the spreadsheet were rechecked for accuracy. Calculation of total score and mean were also rechecked on random assessments. Assessment reports were skimmed for mention of sensory or motor handicaps, or severe behavior problems. Residential unit at date of administration was recorded on the data form.

Phase Two

A master list containing ID number, subject name, birthdate, visual handicap, as well as functioning level and residential unit at initial assessment was prepared. From computer printouts of the institutional population, DS and non-DS subjects were matched 1:1 as previously described in

the subjects section. Non-DS subjects were assigned ID numbers to preserve anonymity.

Demographic data for all non-DS subjects was drawn from computer printouts and recorded on the data form. Test scores for all Pyramid Scales administrations were drawn from cumulative records of psychological assessment reports. Total score and mean per administration were calculated and all information entered on the data form. Non-DS subjects were assigned to 1 of 4 subgroups based on age and functioning level at initial administration, and a recheck of DS/non-DS matches conducted.

Phase Three

Subjects with a 1985-86 measure on The Pyramid Scales eligible for a 3-4 year retest, but who had not been reassessed by Psychological Services were targetted. Retest data on 30 of 49 possible subjects was collected from Pyramid Scale protocols completed by residential direct care staff under the supervision of a chartered psychologist attached to a cluster of residential units. Preliminary inspection of the data indicated incomplete scoring on protocols, or great fluctuation in total score and mean from initial administrations conducted by Psychological Services assessors. Supervising psychologists also warned of inaccuracies in calculation of subdomain percentages. A decision was made to exclude non-Psychological Services data therefore. This study was subsequently limited to use of 3

to 4 year retest data collected by the Psychological Services Assessment Team. Although retest data existed for 56 of 140 DS subjects (40%), and for 64 of 140 non-DS subjects (46%), these subjects were not matched pairs.

All relevant data recorded on the data forms were keypunched and entered into the University of Alberta's computer system for statistical analysis.

Data Analysis

Descriptive statistics (i.e., n, M, SD, and percentage) to summarize sample groups on gender, years institutionalized, and chronological age and functioning level at assessment were computed using the SPSS^x statistical package.

Data analyses were arranged to answer the research question:

> Do adaptive skills in Down syndrome adults show a greater decline with age than adaptive skills of non-Down syndrome mentally handicapped adults?

Theory underlying this study indicated that DS adults over 35 years of age carry the neuropathology of Alzheimer's disease. The incidence of decline in adaptive skills in older DS adults as a result of suspected neuropathology varies widely in the literature however. Inconclusiveness of past research necessitated an exploratory approach in this study. The research objective was to use exploratory data analysis procedures (Tukey,1977, Erickson & Nosanchuk,

1979) to generate possible hypotheses about the data. Confirmatory data analysis procedures using inferential statistics were to follow to assess the statistical significance of observed differences. The research question was examined in 3 phases.

Cross-sectional Exploratory Analysis

Cross-sectional data were examined to determine if differences in subgroups existed with age. Exploratory data analysis was conducted on percentage competency scores across 20 subdomains and total score on the Pyramid Scales. For each subgroup, median, 75th percentile, 25th percentile, high score and low score were computed per subdomain. Schematic plots of median (Md), interquartile range (IQR), outlier and extreme outlier scores were graphed using the MANOVA function from the SPSS^{x} statistical package. Visual analysis focused on examination of the level (Md) and spread (IQR) of subdomain scores across etiology, age, and functioning level. Patterns present in the data were noted. Scores deviating greatly from the midspread were targetted for investigation because of the possible effect of neuropathology on behavior. To examine outlier and extreme outlier scores, stem and leaf displays were plotted for all subgroups across all subdomains. Each outlying score was identified by subject, investigated as a valid score, and a decision was made to retain or remove the score from future analyses. All invalid outlier scores per subdomain were

removed from both initial assessment and retest data. Numerical summaries for all subgroups across all subdomains were recomputed and charted for visual analysis. Trends in the data were identified for further statistical analysis.

Cross-sectional Confirmatory Analysis

Confirmatory data analysis formed the second phase of analysis. Five adaptive skill domains (Sensory-Motor, Self-Help, Communication, Socialization, and Cognitive) were formed from aggregations of related subdomains. Domain scores were used because (1) they provide an overall measure of the level of adaptive behavior on related skills, and (2) their use helps to reduce the possibility of a Type I error because only 5 statistical tests are computed rather than 20 required for testing subdomains separately.

A univariate (2 x 2 x 2) three way analysis of variance was computed using the SPSS^x statistical package. Main effects and interaction across etiology (DS and non-DS), age (<40 years and 40+ years), and functioning level (mild/moderate and severe/profound) were examined. Dependent variables were 5 adaptive behavior domains. A .05 level of significance was used for these statistical tests. The purpose in performing these statistical tests was to determine if significant differences in etiology, age, or functioning level existed between subgroups.

Longitudinal Analysis

A longitudinal comparison was subsequently conducted to assess change in adaptive skills with age in DS adults and their non-DS agemates. Data gathered on subjects following a 3 or 4 year retest interval were analyzed. Examination of subdomain items assessed on the Pyramid Scales revealed that, on average, a 5 point decline in subdomain score would alter the manner in which caregivers interact with an individual (i.e., additional prompts, physical assistance in activities of daily living, or alteration in day programs would be required). Clinical significance was established as a minimum change of 100 points on the total score, or the mean of 20 subdomains ±5 points.

Subdomains recording clinically significant changes in mean score at retest were identified by etiology and age group. Repeated measures 2 way ANOVA's (age x time of assessment) were calculated to assess statistical significance within each etiology.

Frequencies of individuals who recorded a clinically significant change on the mean score were tabulated. Chi-square statistics to test the association between etiology and direction of behavior change by age group at mean score ±5 points were computed using the CROSSTABULATION function of the SPSS^x package. A .05 level of significance was used. Individual profiles were examined to determine if patterns of adaptive behavior change existed within etiologies.

Cumulative files containing caregiver reports prepared for annual Individual Program Plan (IPP) conferences were reviewed for 4 DS adults who died prior to data analysis, and for whom neuropathological results were available. The timing and the sequence of adaptive skill decline were noted.

CHAPTER IV

Results

Findings in this study are summarized systematically. Results gained through cross-sectional comparison of DS and non-DS groups are presented first. Findings gained through longitudinal analysis follow as a secondary component of the study.

The overall pattern of results gained from analysis of cross-sectional data indicated that no significant age-related differences in adaptive skills existed between mentally handicapped adults with and without Down syndrome. As expected, significant differences were reported as a function of level of retardation. Conversely, longitudinal analysis indicated that a greater number of Down syndrome adults recorded a decline in adaptive skills with age than their non-Down syndrome peers. This was demonstrated primarily in DS persons over 40 years of age. Clinically significant declines in Washing/Grooming, Gross Motor, Receptive Language, Social Interaction, Dressing, Domestic Behavior, and Toileting subdomains were recorded at retest in the over 40 DS group. The progression of decline in 4 DS persons with confirmed AD at post-mortem demonstrated a similar pattern, although the age at onset and rate of decline varied per individual.

Cross-sectional Analysis

Median Analysis

During the first phase of analysis, competence in adaptive skills as a function of etiology, age, and functioning level was examined by visually comparing midspreads of subgroups across 20 subdomains and total score. Results on each subdomain were remarkably consistent for subgroups across both etiology and age. Generally, median scores varied by only one or two items (i.e., 3% to 6%) per Pyramid Scale subdomain.

By etiology. Examination across etiology revealed that Down syndrome subgroups scored equal to or above their peers on 79.8% of subdomains (67 of 84 comparisons). The greatest discrepancy between median scores was seen among subdomains in the Socialization domain. The under 40 year-old, mild/moderate (m/m) DS subgroup surpassed their agemates in Recreation/Leisure by 15%, Domestic Behavior by 25%, and Vocational subdomains by 11%. The non-DS group with its variety of handicapping conditions, was a less homogeneous cluster than the DS group. No consistent trend in variability between groups was demonstrated however. Greater variability in the 40+ m/m non-DS subgroup in Gross Motor and Domestic Behavior subdomains was attributed to a cluster of persons confined to wheelchairs.

By age. Examination of differences with age within the DS group indicated that for the over 40 year old

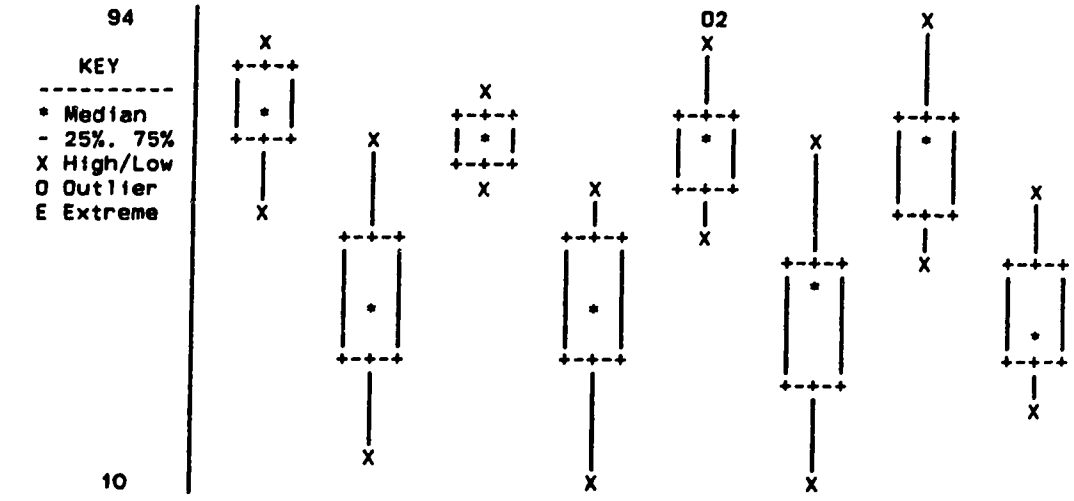
mild/moderate subgroup, decreased competence was recorded in the following subdomains: Numbers by 13%, Money by 12%, Writing by 10%, Visual Responsiveness by 9%, and Social Interaction by 9%. The older severe/profound DS subgroup recorded less competence in Auditory Responsiveness by 9%. Conversely, stability of median scores across age was consistently noted in the non-DS group. Severe/profound subgroups recorded consistently less competence across subdomains than those with mild/moderate retardation. Median and interquartile range for all subgroups per subdomain are presented in Table A (see Appendix C). Box plots per subgroup on the mean of 20 subdomains, graphed in Figure 1, demonstrates the uniformity across distributions typically seen in the data.

Resistance to extreme scores are a feature of midspread analysis and help isolate patterns present in the data. In this study, the effect of extreme scores as possible indicators of the impact of neuropathology on adaptive behavior, necessitated their inclusion in subsequent data analyses. Mean and standard deviation statistics were therefore used for remaining comparisons. In addition, the 20 subdomains were aggregated to 5 domains for further analyses.

Means Analysis

By domain. Visual comparison of means for 8 subgroups across domains addressed the question of the effect of

Figure 1 Box Plots of Percentage Competency on the Mean of 20 Subdomains per Subgroup



Subgroup:								
	1	DS	<40	m/m	5	non-DS	<40	m/m
	2	DS	<40	s/p	6	non-DS	<40	s/p
	3	DS	40+	m/m	7	non-DS	40+	m/m
	4	DS	40+	s/p	8	non-DS	40+	s/p

etiology, age, and functioning level on adaptive behavior. Results presented in Table B (see Appendix C) indicate that DS subgroups under 40 years of age equal or slightly surpass their peers in adaptive skill competence on all domains except Communication. DS subgroups over 40 years of age recorded slightly higher levels of competence than their agemates in all domains except in the Cognitive domain for the mild/moderate classification, and in the Sensory-Motor and Communication domains for those with a severe/profound level of functioning. For all severe/profound functioning subgroups, scores approached a floor in the Cognitive domain however. Interestingly, with age, mild/moderate DS subgroups recorded a decline of 10.4% in the Cognitive domain.

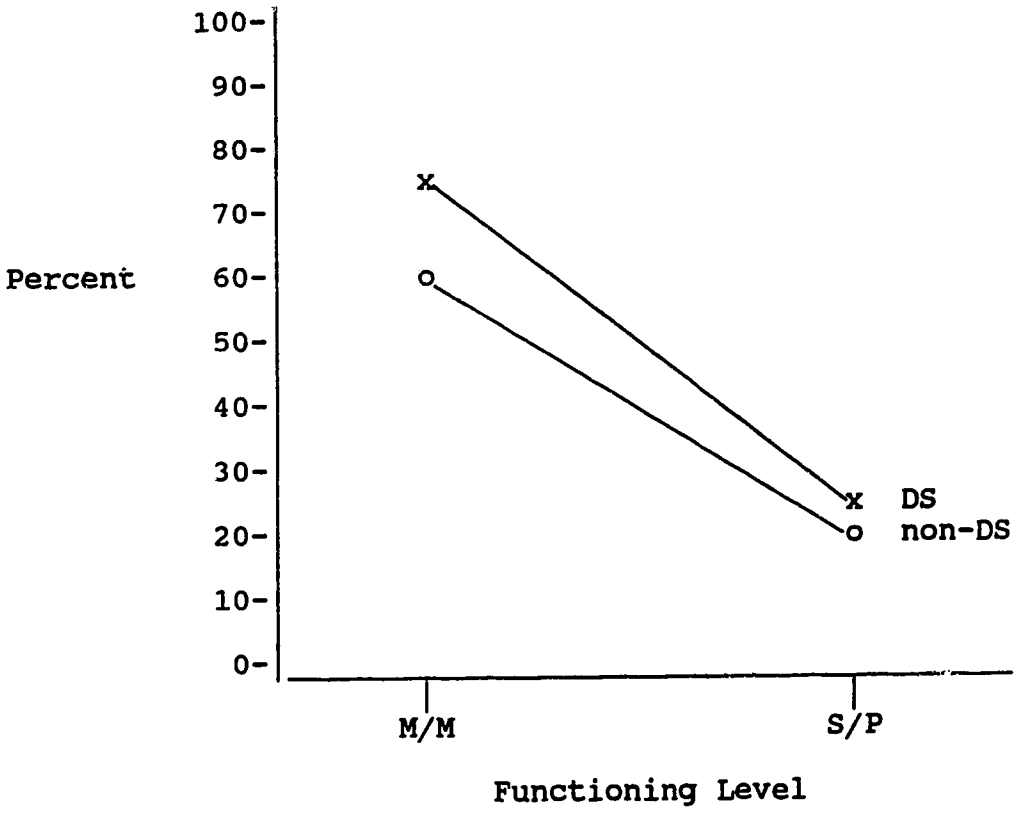
A univariate three way analysis of variance (etiology x age x functioning level) confirmed statistically significant differences for functioning level across all domains: Sensory-Motor ($F = 171.31$), Self-Help ($F = 190.64$), Communication ($F = 260.84$), Socialization ($F = 534.66$), Cognitive ($F = 386.42$) at $df(1,272)$ and $p < .001$ indicating as expected, that mild/moderate subgroups consistently display more competence in adaptive behavior. Main effects of etiology on Self-Help [$F(1,272) = 5.28$, $p < .022$], and Socialization domains [$F(1,272) = 9.32$, $p < .002$] confirmed greater competence in self-care and social skills for DS persons compared to their non-DS peers. The significant

interaction of etiology and functioning level on the Socialization domain [$F(1,272) = 6.96$, $p < .009$] indicated that m/m DS subjects have greater social skills than their non-DS peers, but that persons with s/p retardation do not show the same group differences. Figure 2 illustrates the interaction effect of etiology and functioning level. Statistically significant differences as a function of age were not reported via cross-sectional analysis.

By subdomain. Means across individual subdomains for all subgroups are presented for visual comparison in Table C (see Appendix C). DS subgroups recorded higher levels of competence on 71.4% of subdomains (60 of 84 comparisons). In 7 subdomains valid low scores contributed to the slight decline of DS subgroups to below competence levels of their peers previously recorded by median analysis.

Minimal differences between subgroups were reported on Sensory-Motor skills where both mild/moderate and severe/profound DS groups over 40 years of age displayed less competence than peers in auditory responsiveness (M = 89.7 DS, and M = 92.4 non-DS m/m), (M = 55.7 DS, and M = 74.8 non-DS s/p); and visual responsiveness (M = 87.8 DS, and M = 90.0 non-DS m/m), (M = 53.8 DS, and M = 57.5 non-DS s/p). Those in the severe/profound classification recorded slightly lower scores in visual responsiveness (M = 55.7 DS, and M = 58.7 non-DS); and tactile responsiveness (M = 83.9 DS, and M = 85.3 non-DS) in younger DS groups but added less

Figure 2 Mean Percentage Competency as a Function of Etiology by Functioning Level Interaction on the Socialization Domain



competence in auditory responsiveness (M = 65.7 DS, and M = 74.8 non-DS) and fine motor skills (M = 58.4 DS, and M = 60.8 non-DS) to this with age.

Cognitive skills including reading (M = 7.1 DS, and M = 8.4 non-DS), writing (M = 17.9 DS, and M = 19.5 non-DS), and number skills (M = 2.8 DS, and M = 4.4 non-DS), which were poorly developed in all subjects in the under 40 year old severe/profound DS subgroup, displayed a floor effect. Although the younger DS mild/moderate subgroup initially recorded higher competence levels than comparable agemates in the cognitive area, with age less competence was demonstrated in reading (M = 22.0 DS, and M = 30.7 non-DS), writing (M = 46.0 DS, and M = 55.6 non-DS), number (M = 24.6 DS, and M = 30.2 non-DS), time (M = 43.9 DS, and M = 50.3 non-DS), and money skills (M = 22.4 DS, and M = 28.0 non-DS). Comparison within the DS group of younger and older subgroups revealed less competence with age in Numbers (12.6%), Reading (11%), Money (10.6%), Writing (9.5%), and Time (9.2%) subdomains.

Slightly poorer performance in communication skills was recorded among both younger and older DS subgroups in receptive language, accompanying deficits in auditory and visual responsiveness. Interestingly however, with age the DS mild/moderate subgroup maintained expressive language skills to slightly surpass their peers.

Eating ($\underline{M}$ = 69.2 DS, and $\underline{M}$ = 71.1 non-DS) was the only Self-Help subdomain to record less competence with age, and this minimal change occurred in the DS severe/profound grouping where deficits in tactile and visual responsiveness, and fine motor skills were also recorded with age. Due to the minimal change recorded across subdomains, and the likelihood of a large number of comparisons producing significant results by chance, tests of statistical significance were not conducted per subdomain. Visual comparison of means across subdomains served only to indicate which skill areas were influenced by valid low scores.

Cross-sectional analysis was designed to focus on age-related differences in adaptive behavior across subgroups at one particular point in time. As this methodology was unable to address age-related changes in adaptive behavior, longitudinal analysis was conducted to examine adaptive behavior within the same individuals at two different points in time.

Longitudinal Analysis

Data from DS individuals retested after a 3 to 4 year interval ($\underline{N}$ = 56) were examined to assess the effects of age on adaptive behavior, and to see if trends in the direction of behavior change occurred with age. Comparisons were also made within the non-DS group ($\underline{N}$ = 64) to determine if similar patterns existed regardless of etiology.

Demographic characteristics of DS and non-DS groups at retest are presented in Table 1 (see page 46).

Results obtained through longitudinal analysis were generally consistent with that observed via cross-sectional examination of distinct groups of different ages. Stability of behavior regardless of age or etiology was the most predominant finding. However, longitudinal data indicated that beyond age 40 a number of individuals in the DS group began to show a trend toward decreased stability in their general level of adaptive behavior, and more specifically, toward declines in the Self-Help and Communication domains. Further analysis indicated that for those beyond age 40, the greatest declines occurred in washing/grooming, gross motor, receptive language, social interaction, dressing, domestic behavior, and toileting skills.

Change in Adaptive Behavior

By etiology. Initially, mean scores for each of 20 subdomains and overall assessment mean for the total group were plotted on initial assessment (pretest), and on retest data. Although they were not matched groups, results revealed that virtually identical profiles existed for DS and non-DS groups. Generally, slightly lower levels of competence were recorded by the DS group per subdomain. Because few young mild/moderate DS persons were retested (n = 4), this may be attributed to a predominance of scores from individuals in the severe/profound category. At retest

both DS and non-DS groups showed remarkable consistency to their respective pretest means.

An emerging trend was noted within the DS profile however. Subdomains which recorded a clinically significant change (i.e., mean ± 5%) were: Washing/Grooming (decline = 7.5%), Dressing (decline = 5.8%), and Receptive Language (decline = 5.1%). A clinically significant decline of 5.1% was reported on the Vocational subdomain for both DS and non-DS groups, while increases of 5.5% in the DS group, and 4.4% in the non-DS group were recorded on the Visual Responsiveness subdomain. Within the non-DS group an increase of 4.7% on the Social Interaction subdomain approached clinical significance. Profiles at pretest and retest are presented for the DS group in Figure 3, and the non-DS group in Figure 4. Percentage competency per subdomain for each group at pretest, at retest, and change score are presented in Table 2.

By age. DS and non-DS groups were subsequently divided into subgroups, those under 40 or over 40 years of age at pretest. Percentage change per subdomain at retest was then examined separately by etiology. Results indicated that for the DS group, stability of adaptive behavior across time was predominant in the under 40 year old subgroup. Only on Vocational and Dressing subdomains were clinically significant declines recorded (-5.66%, and -5.05% respectively). Interestingly, the over 40 year old DS

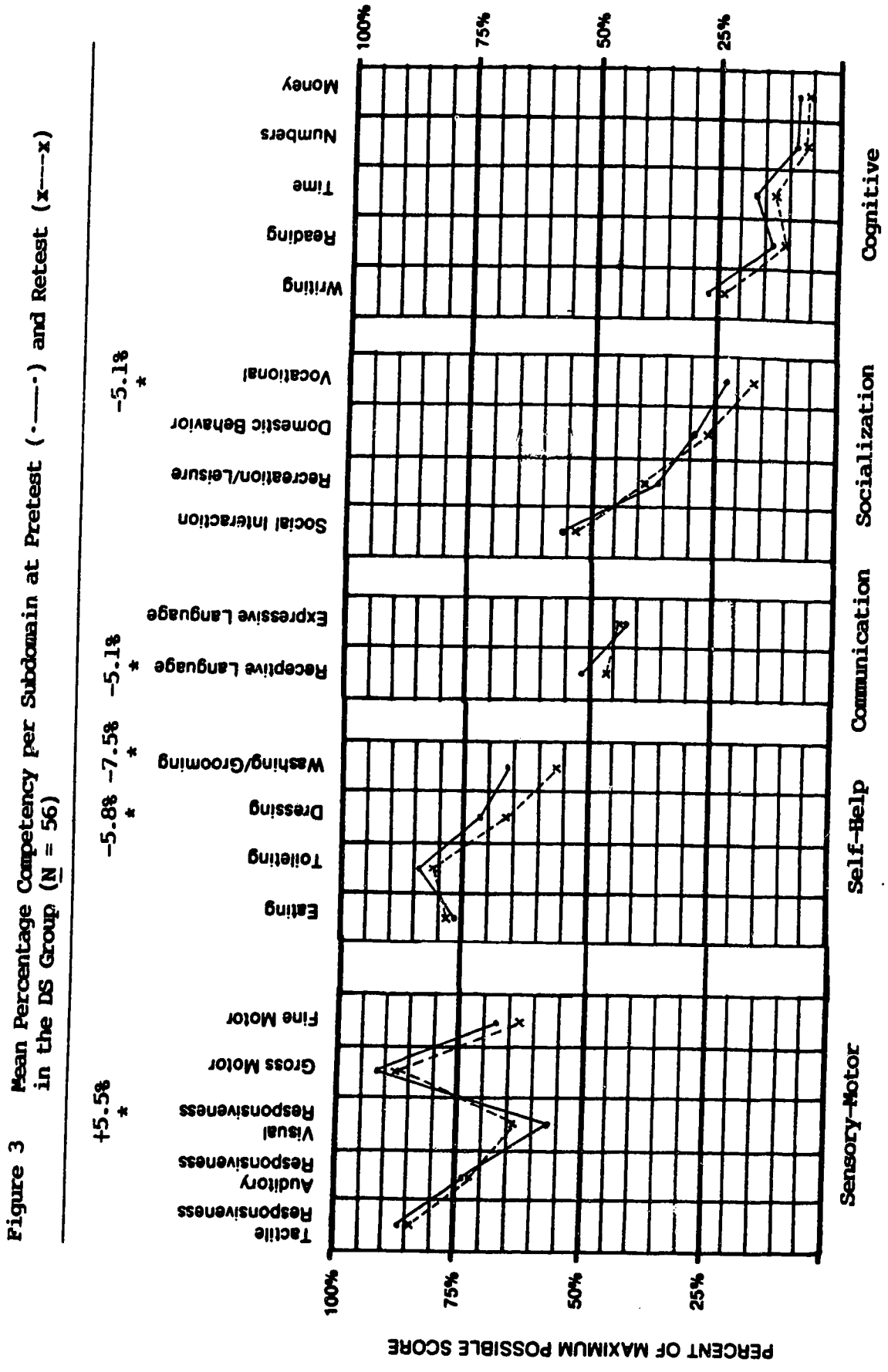


Figure 3 Mean Percentage Competency per Subdomain at Pretest (·——·) and Retest (x—x) in the DS Group ($\underline{N}$ = 56)

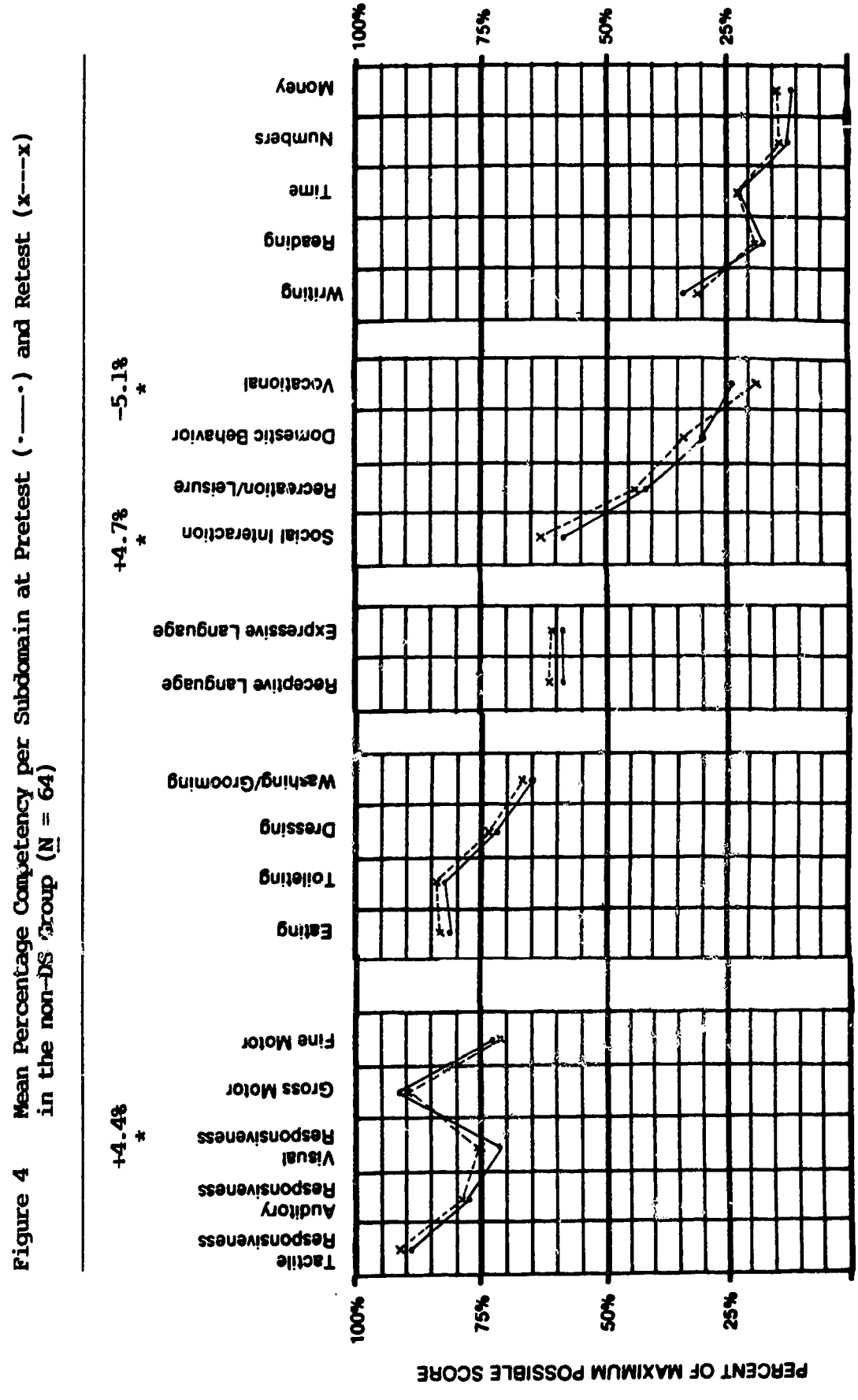


Figure 4 Mean Percentage Competency per Subdomain at Pretest (•——•) and Retest (x---x) in the non-DS Group (N = 64)

Table 2 Mean Percentage Competency per Subdomain at Pretest and Retest as a Function of Etiology

	DS (N = 56)			non-DS (N = 64)		
Subdomain	Pretest	Retest	% Change	Pretest	Retest	% Change
Sensory-Motor						
TR	86.12	85.55	-0.57	89.47	90.65	+1.18
AR	73.55	73.01	-0.54	77.46	79.18	+1.72
VR	57.73	63.19	+5.46 *	70.56	74.96	+4.40
GMOT	90.57	86.98	-3.59	90.00	90.09	+0.09
FMOT	66.23	63.80	-2.43	72.28	70.93	-1.35
Self-Help						
EAT	75.82	76.41	+0.59	80.79	81.65	+0.86
TOIL	83.44	81.98	-1.46	83.87	84.15	+0.28
DRES	71.91	66.14	-5.77 *	71.90	74.20	+2.30
WASH	65.10	57.62	-7.48 *	65.37	65.00	-0.37
Communication						
RLAN	51.25	46.16	-5.09 *	59.04	61.35	+2.31
ELAN	43.26	43.35	+0.09	59.68	59.79	+0.11
Socialization						
SOCI	54.96	53.26	-1.70	57.95	62.62	+4.67
REC	36.33	39.26	+2.93	40.95	43.03	+2.08
DOME	28.91	25.85	-3.06	29.68	32.93	+3.25
VOC	22.53	17.37	-5.16 *	23.31	18.25	-5.06 *
Cognitive						
WRIT	27.07	24.89	-2.18	32.89	30.68	-2.21
READ	12.76	11.26	-1.50	16.00	16.54	+0.54
TIME	15.85	13.51	-2.34	21.25	21.73	+0.48
NUM	8.53	6.73	-1.80	12.84	13.35	+0.51
MON	7.82	7.25	-0.57	11.12	12.89	+1.77
Mean Score	48.91	46.96	-1.95	53.35	54.15	+0.80

*Clinically significant change = mean $\pm$ 5%

subgroup recorded clinically significant declines at retest in the following subdomains: Washing/Grooming (-14.00%), Gross Motor (-8.95%), Receptive Language (-8.62%), Social Interaction (-7.67%), Dressing (-6.96%), Domestic Behavior (-6.81%), and Toileting (-5.05%). These findings are particularly important as declines of this magnitude require additional prompting and physical assistance to aging persons with DS, thereby altering caregiver interactions. Only on the Visual Responsiveness subdomain did a clinically significant increase in competency (+7.85%) occur at retest, possibly attributed to less stringent adherence to assessment criteria since both age groups across both etiologies recorded an increase on retest. Percentage competency at pretest, retest, and change score per subdomain are presented for DS subgroups in Table 3.

Two way Analyses of Variance with a repeated measures factor (age X time of assessment) were conducted for the DS group. Statistically significant main effects of time of assessment were reported on the following subdomains: Dressing [$F(1,54) = 10.15$, $p<.002$]; Receptive Language [$F(1,54) = 5.64$, $p<.021$]; Vocational [$F(1,54) = 5.19$, $p<.027$]; Domestic Behavior [$F(1,54) = 4.63$, $p<.036$]; Visual Responsiveness [$F(1,54) = 4.46$, $p<.039$]; and on the mean of 20 subdomains [$F(1,54) = 5.40$, $p<.024$]. This indicated that DS subgroups regardless of age displayed less competence in these skill areas on retest, with the exception of increases

Table 3 Mean Percentage Competency per Subdomain at Pretest and Retest as a Function of Age in the DS Group (N = 56)

	<40 Years (n = 35)			40+ Years (n = 21)		
Subdomain	Pretest	Retest	% Change	Pretest	Retest	% Change
Sensory-Motor						
TR	86.97	84.48	-2.49	84.71	87.33	+2.62
AR	73.40	71.80	-1.60	73.81	75.04	+1.23
VR	56.51	60.54	+4.03	59.76	67.61	+7.85 *
GMOT	90.05	89.68	-0.37	91.42	82.47	-8.95 *
FMOT	63.80	62.28	-1.52	70.28	66.33	-3.95
Self-Help						
EAT	76.28	76.28	0.00	75.04	76.61	+1.57
TOIL	80.85	81.54	+0.69	87.76	82.71	-5.05 *
DRES	68.85	63.80	-5.05 *	77.00	70.04	-6.96 *
WASH	58.20	54.62	-3.58	76.61	62.61	-14.00 *
Communication						
RLAN	47.48	44.51	-2.97	57.52	48.90	-8.62 *
ELAN	37.77	37.00	-0.77	52.42	53.95	+1.53
Socialization						
SOCI	49.34	51.22	+1.88	64.33	56.66	-7.67 *
REC	34.71	38.00	+3.29	39.04	41.38	+2.34
DOME	25.08	24.28	-0.80	35.28	28.47	-6.81 *
VOC	20.57	14.91	-5.66 *	25.81	21.47	-4.34
Cognitive						
WRIT	25.91	23.45	-2.46	29.00	27.28	-1.72
READ	13.02	10.97	-2.05	12.33	11.76	-0.57
TIME	14.42	12.11	-2.31	18.23	15.85	-2.38
NUM	7.85	6.31	-1.54	9.66	7.42	-2.24
MON	7.77	6.00	-1.77	7.90	9.33	+1.43
Mean Score	46.94	45.37	-1.57	52.19	49.61	-2.58

*Clinically significant change = mean ± 5%

at retest in Visual Responsiveness. Significant interactions of age by time of assessment reported on Washing/Grooming [$F(1,54) = 6.94$, $p<.011$]; Gross Motor [$F(1,54) = 9.05$, $p<.004$]; and Social Interaction subdomains [$F(1,54) = 4.85$, $p<.032$], demonstrated that the older DS subgroup recorded large declines on retest, but that the younger DS subgroup did not show the same group differences. Age by time of assessment interaction effects are presented in Figure 5. Although measures of statistical significance to confirm the likelihood of results being recorded by chance are useful, it is of greater importance to focus on skill areas where clinically significant changes occur, both as indicators of a possible disease process, and as cues to alter service provision.

Examination of changes in competence at retest in non-DS subgroups under 40, or over 40 years of age were conducted to determine if similar patterns in adaptive behavior existed regardless of etiology. Stability of adaptive skills was recorded across both age groups. Exceptions to this were noted in the under 40 subgroup where a clinically significant decline of 6.02% was recorded in the Vocational subdomain, and in the over 40 subgroup where clinically significant increases of 12.90% in Domestic Behavior; 5.47% in Social Interaction; and 5.00% in Receptive Language subdomains were reported at retest. Percentage competency at pretest, retest, and change score

Figure 5 Mean Percentage Competency as a Function of Age by Time of Assessment Interaction for the DS Group

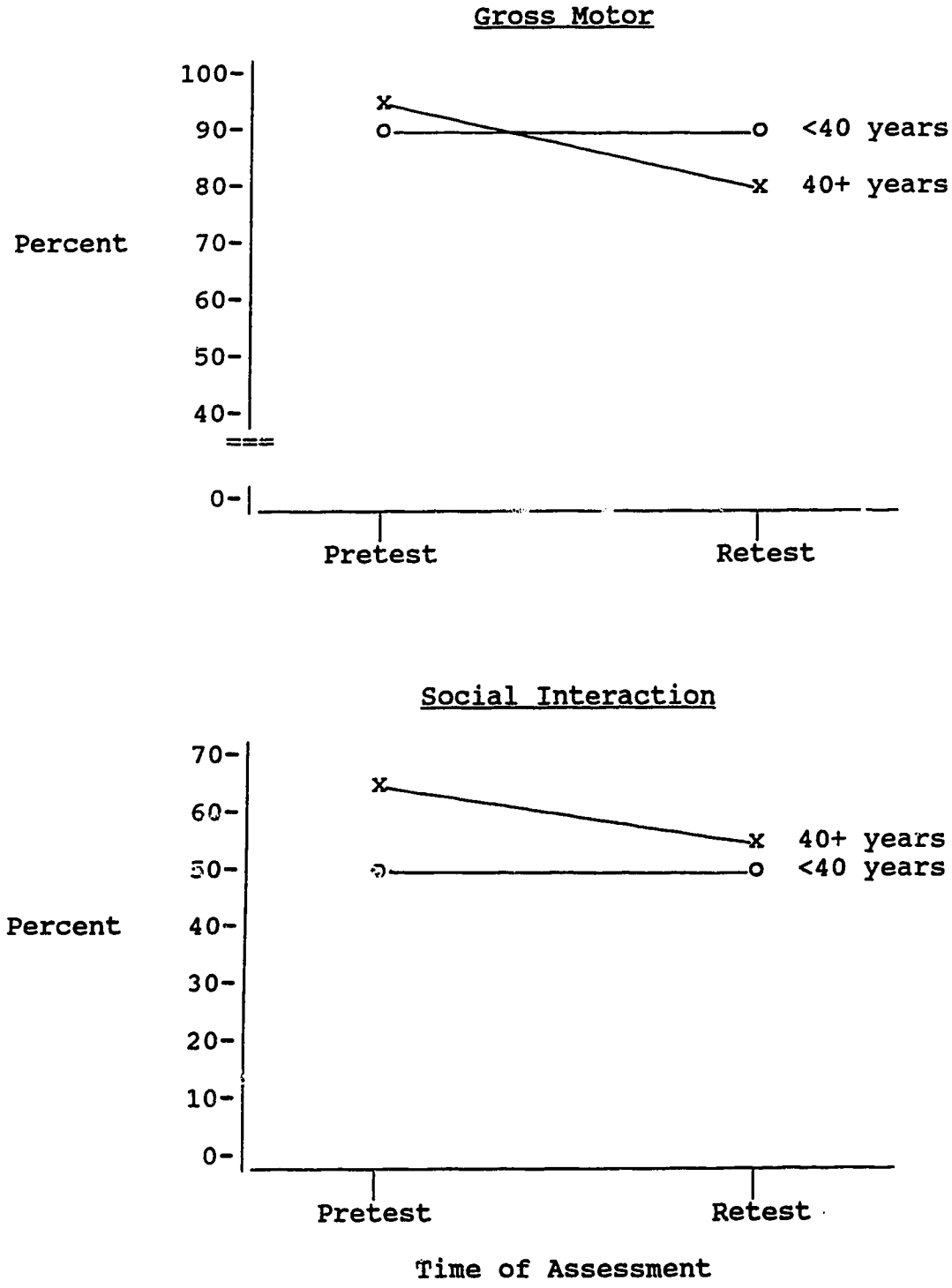
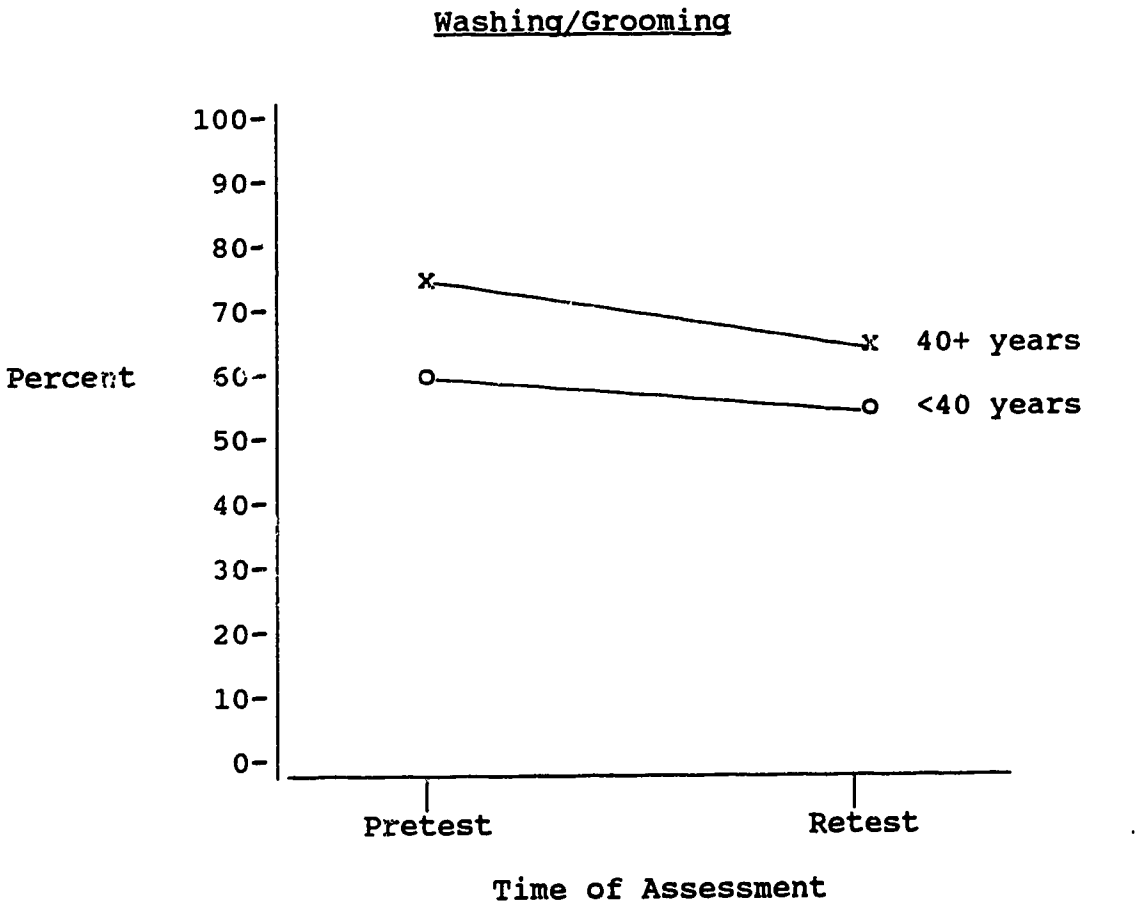


Figure 5 Continued



per subdomain are presented for non-DS subgroups in Table 4.

Repeated measures ANOVA's (age X time of assessment) calculated for the non-DS group reported statistical significance for main effects of time of assessment on: Visual Responsiveness [$F(1,62) = 5.37$, $p<.024$]; Dressing [$F(1,62) = 5.11$, $p<.027$]; and Social Interaction [$F(1,62) = 4.94$, $p<.03$] subdomains demonstrating that increased competence at retest in the non-DS group occurred regardless of age. Analysis of variance yielded significant interactions of age by time of assessment for Domestic Behavior [$F(1,62) = 9.26$, $p<.003$]; and Time [$F(1,62) = 3.88$, $p<.05$] indicating that the older non-DS subgroup reported increased competence on retest but that the younger age group did not show the same pattern. Age by time of assessment interaction effects are illustrated in Figure 6.

Change by Number of Individuals

Subsequently, the focus of analysis shifted from change scores per adaptive behavior, to direction of change recorded by a majority of persons. Results suggested that a greater number of DS persons recorded clinically significant declines after age 40.

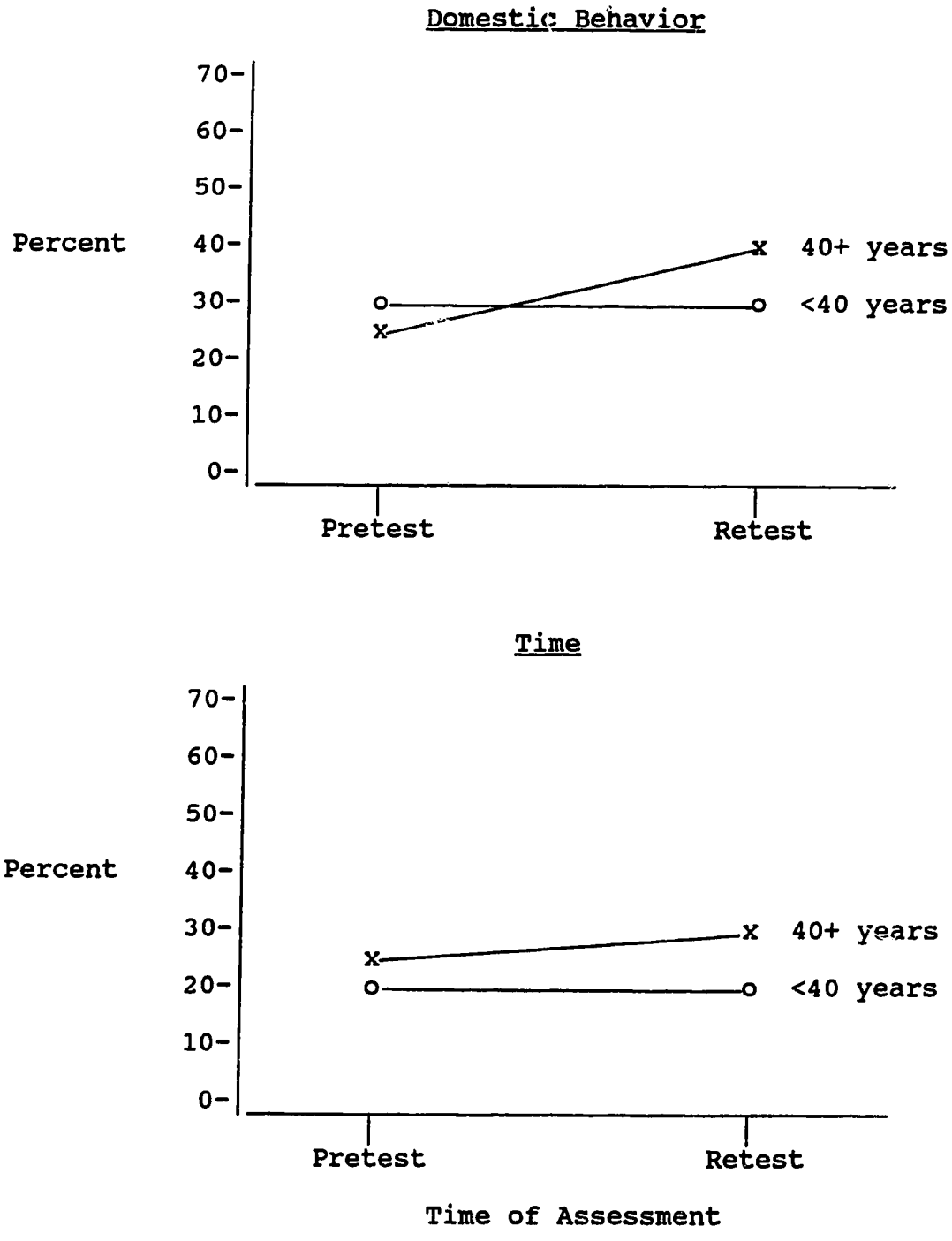
General level of adaptive behavior. Percentages of individuals who recorded clinically significant declines, no change, or gains on the mean score of 20 subdomains were tabulated across groups collapsed on functioning level. Preliminary examination revealed that a similar pattern was

Table 4 Mean Percentage Competency per Subdomain at Pretest and Retest as a Function of Age in the non-DS Group (N = 64)

	<40 Years (n = 45)			40+ Years (n = 19)		
Subdomain	Pretest	Retest	% Change	Pretest	Retest	% Change
Sensory-Motor						
TR	88.40	89.48	+1.08	92.00	93.42	+1.42
AR	78.42	79.51	+1.09	75.21	78.42	+3.21
VR	69.62	74.15	+4.53	72.78	76.89	+4.11
GMOT	91.53	91.46	-0.07	86.36	86.84	+0.48
FMOT	71.02	69.37	-1.65	75.26	74.63	-0.63
Self-Help						
EAT	79.91	80.88	+0.97	82.89	83.47	+0.58
TOIL	83.46	83.62	+0.16	84.84	85.42	+0.58
DRES	70.77	72.17	+1.40	74.57	79.00	+4.43
WASH	62.35	62.08	-0.27	72.52	71.89	-0.63
Communication						
RLAN	59.06	60.24	+1.18	59.00	64.00	+5.00 *
ELAN	58.11	57.26	-0.85	63.42	65.78	+2.36
Socialization						
SOCI	56.55	60.88	+4.33	61.26	66.73	+5.47 *
REC	39.75	41.97	+2.71	43.78	45.52	+1.74
DOME	30.26	29.44	-0.82	28.31	41.21	+12.90 *
VOC	23.37	17.35	-6.02 *	23.15	20.36	-2.79
Cognitive						
WRIT	31.37	29.04	-2.33	36.47	34.57	-1.90
READ	14.86	15.24	+0.38	18.68	19.63	+0.95
TIME	19.22	17.91	-1.31	26.05	30.78	+4.73
NUM	11.28	11.68	+0.40	16.52	17.31	+0.79
MON	9.17	10.80	+1.63	15.73	17.04	+2.11
Mean Score	52.44	52.64	+0.20	55.52	57.73	+2.21

*Clinically significant change = mean ± 5%

Figure 6 Mean Percentage Competency as a Function of Age by Time of Assessment Interaction for the non-DS Group



present in both mild/moderate and severe/profound classifications. Results indicated that for the total group collapsed by age (N = 120), the majority of both DS and non-DS persons recorded no clinically significant change in mean score (42.9% and 64.1% respectively). This pattern was replicated in groups under 40 years of age (N = 80) for 48.6% of DS persons, and 60.0% of non-DS subjects. In the over 40 age group (N = 40) however, 42.9% of DS individuals recorded a clinically significant decline, while 5.3% of non-DS persons declined, in contrast to 73.7% of non-DS adults who recorded no change in mean score. This indicates that as DS persons surpass 40 years of age, a greater number demonstrate decreasing stability in their general level of adaptive behavior to record either an increase or decrease in skill competence with age. Conversely, a greater majority of non-DS persons display increasing stability in their general level of adaptive functioning beyond age 40. These results are presented in Figure 7. Chi square analysis confirmed that a significant relationship existed between etiology and direction of change on the mean score, for the total group [$X^2(2) = 10.08$, $p<.007$], and for the over 40 age group [$X^2(2) = 8.77$, $p<.013$]. A significant relationship was not demonstrated in the under 40 age group [$X^2(2) = 3.94$, $p<.139$].

By domain. Examination of adaptive behavior change across domains revealed that Self-Help skills were subject

Figure 7 Direction of Change on the Mean of 20 Subdomains as a Function of Etiology

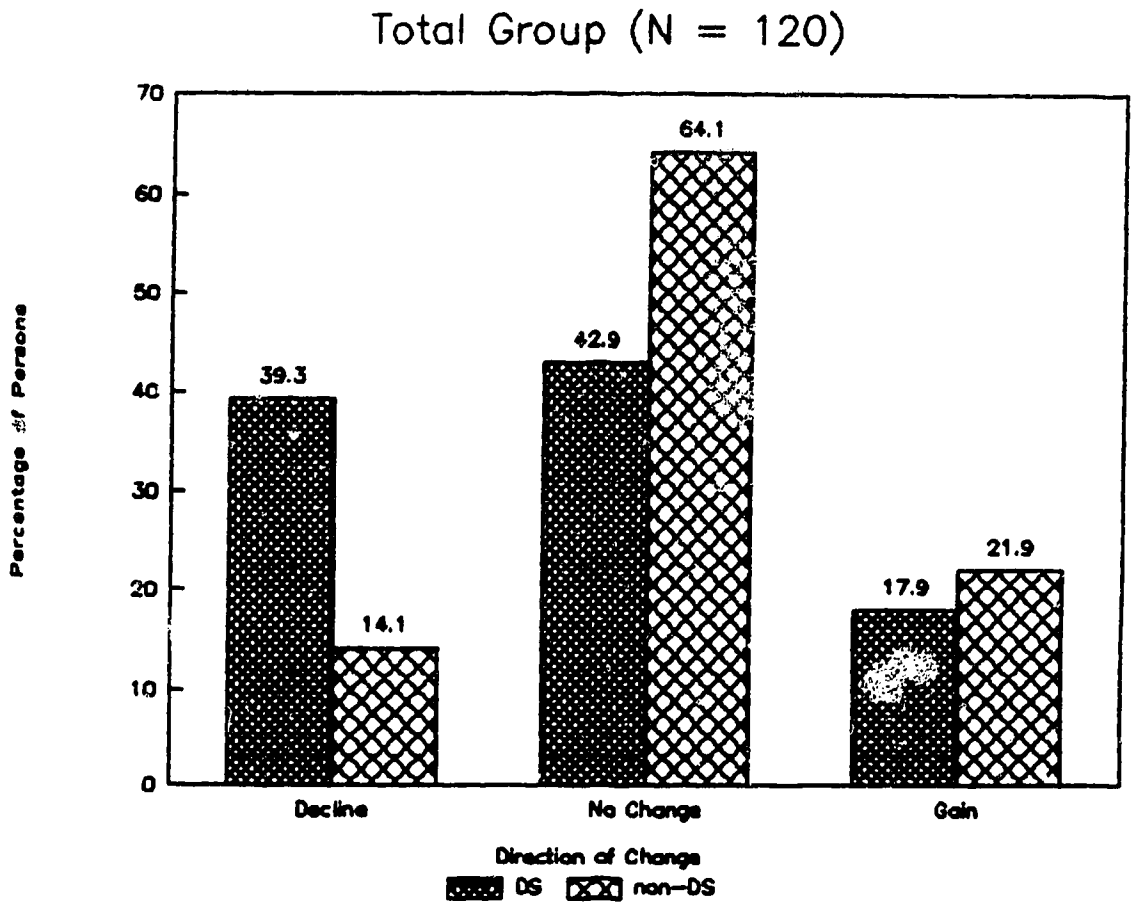
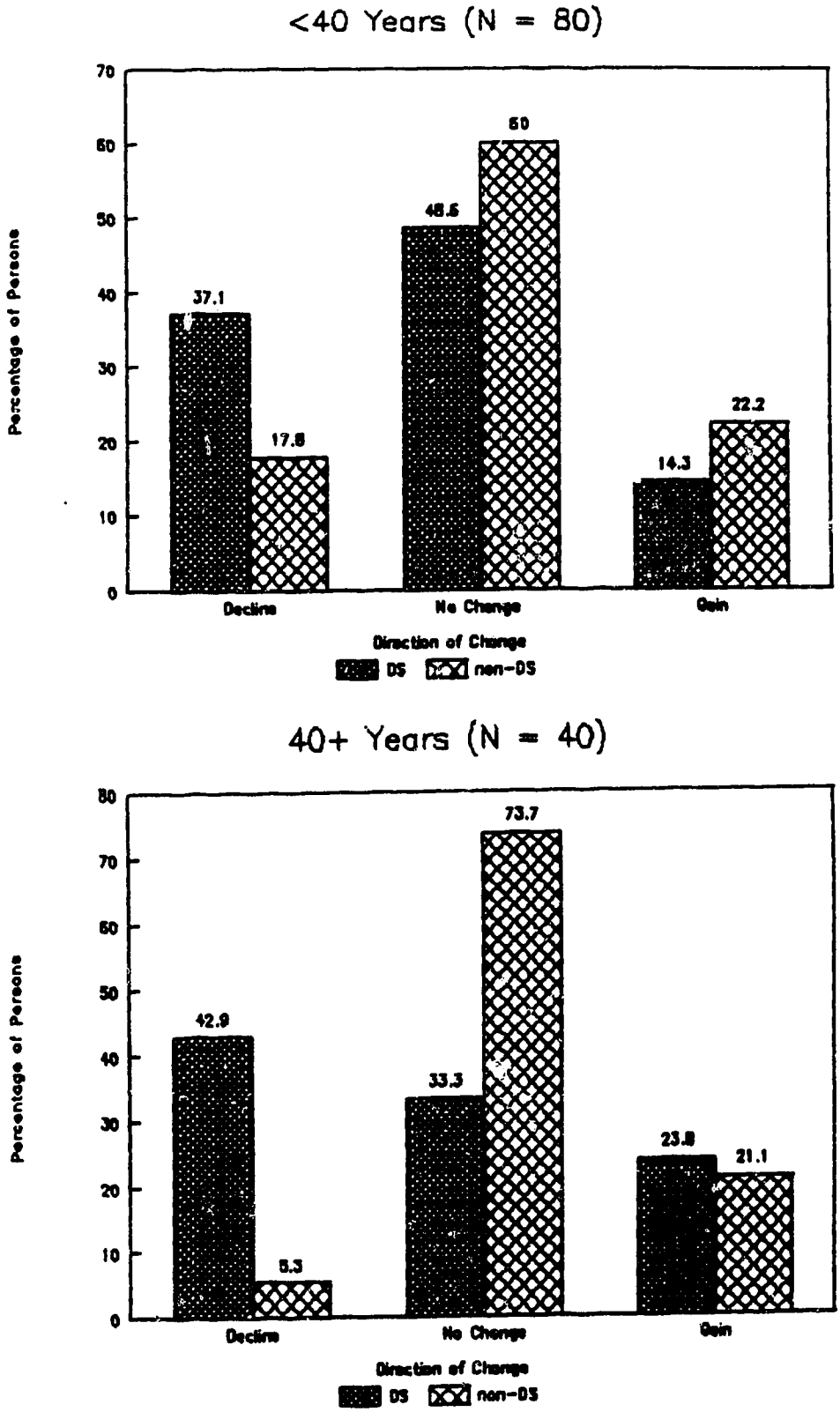


Figure 7 Continued



to decline with age for the majority of DS persons over 40 years. Age did not affect the number of non-DS persons to record stability or change in self-help skills however. For the total group, 42.9% of DS persons recorded a decline, and 42.9% recorded no change on the Self-Help domain, compared to 56.3% of their non-DS peers who maintained skills on this domain. A breakdown by age revealed that in the under 40 age group, 48.6% of DS and 55.6% of non-DS persons recorded no clinically significant change. In the over 40 age group, performance in Self-Help skills declined in 57.1% of DS persons while 57.9% of their non-DS agemates recorded no clinically significant change. The number of non-DS persons to score gains in Self-Help skills remained remarkably consistent regardless of age (26.7% under 40 years, and 26.3% over 40 years). Frequencies of individuals recording changes on the Self-Help domain are presented in Figure 8. Chi square statistics confirmed the association between etiology and direction of change for the total group [$X^2(2) = 9.98$, $p<.007$], and for the over 40 year age group [$X^2(2) = 7.49$, $p<.024$]. Again, a significant difference was not demonstrated in the under 40 age group [$X^2(2) = 3.12$, $p<.209$].

In the Communication domain, clinically and statistically significant declines were reported for DS persons regardless of age. This indicated that DS persons at any age were more likely to decline in communication

Figure 8 Direction of Change on the Self-Help Domain as a Function of Etiology

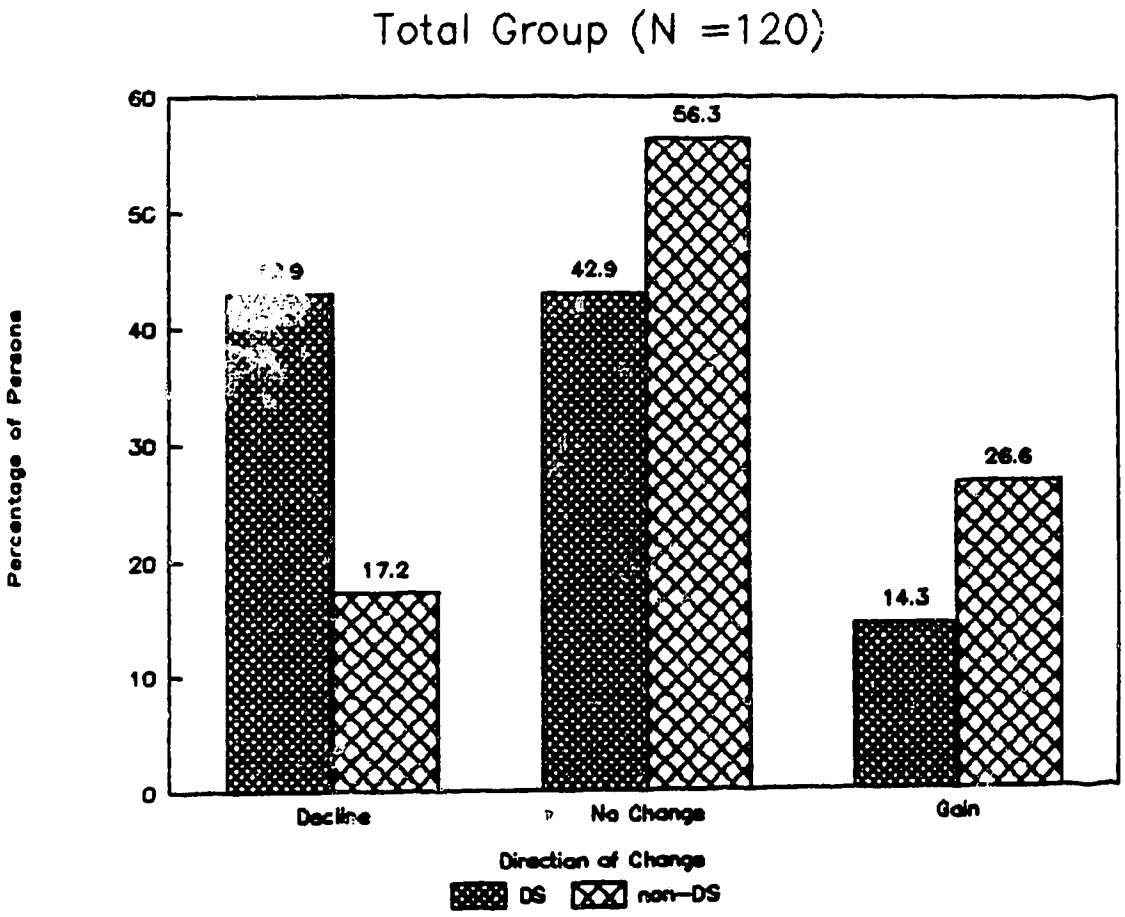
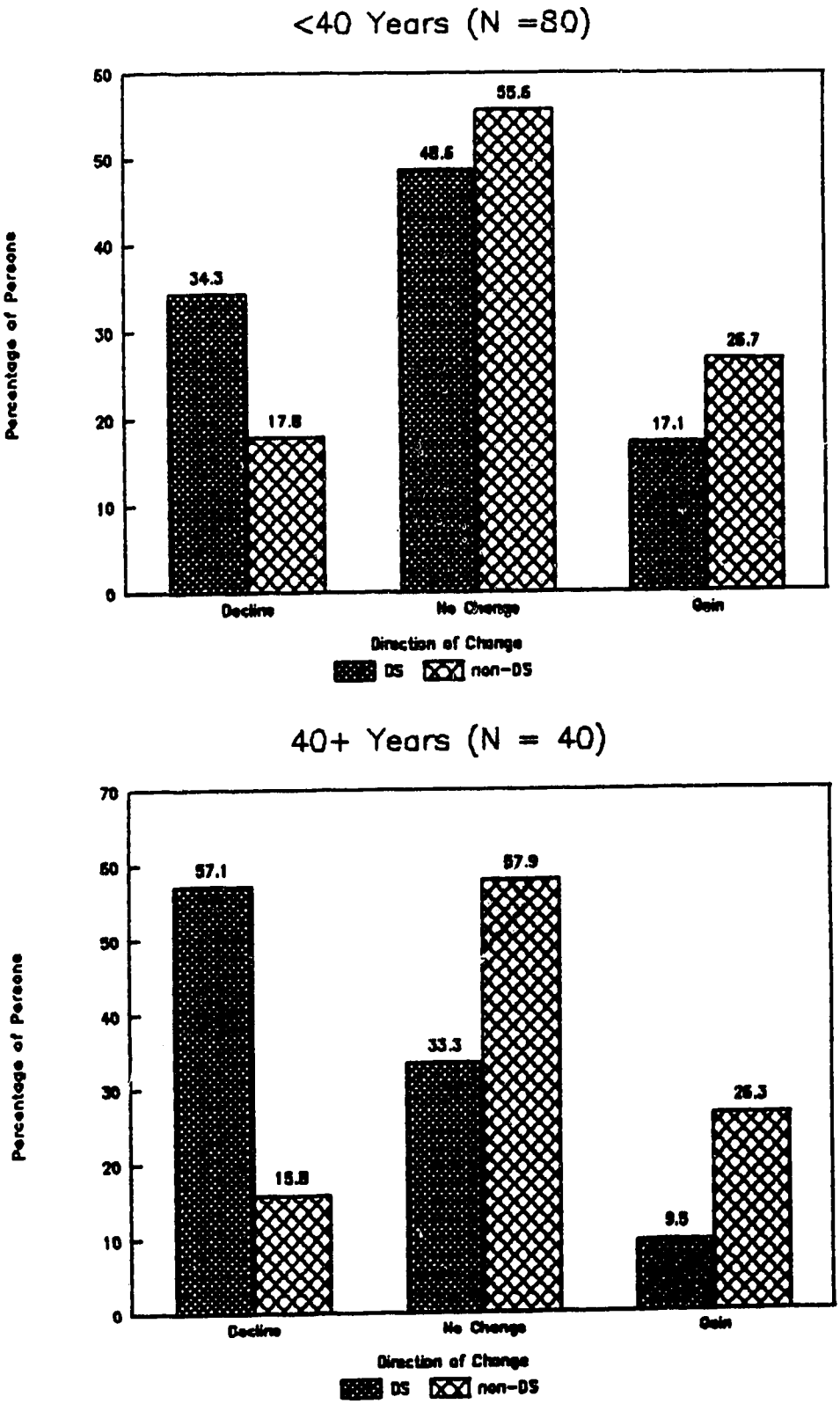


Figure 8 Continued



skills than their non-DS peers, and that this trend became more pronounced beyond age 40. Data showed that for the total group, 50% of DS individuals recorded a decline, while 48.4% of non-DS persons reported no change in domain score. Examination by age group revealed that in the under 40 age group 45.7% of DS adults declined, while 46.7% of non-DS persons recorded no clinically significant change. This trend was replicated and emphasized in the over 40 age group as 57.1% of the DS group declined in communication skills, while 52.6% of their non-DS agemates recorded no change. These results are presented in Figure 9. Chi square analysis confirmed the association of etiology and direction of change in the Communication domain. Statistically significant differences were recorded for the total group [$X^2(2) = 13.83$, $p<.001$]; for the under 40 age group [$X^2(2) = 8.14$, $p<.017$]; and for the over 40 age group [$X^2(2) = 7.39$, $p<.025$].

Chi square statistics tabulated by etiology and age did not demonstrate statistical significance on Sensory-Motor, Socialization, or Cognitive domains. This indicated that observed frequencies did not deviate from that expected by chance.

By subdomain. Subdomains on which an association between direction of behavior change and etiology were reported via chi square analysis were: Dressing [$X^2(2) = 6.32$, $p<.043$], Eating [$X^2(2) = 6.17$, $p<.045$], and

Figure 9 Direction of Change on the Communication Domain as a Function of Etiology

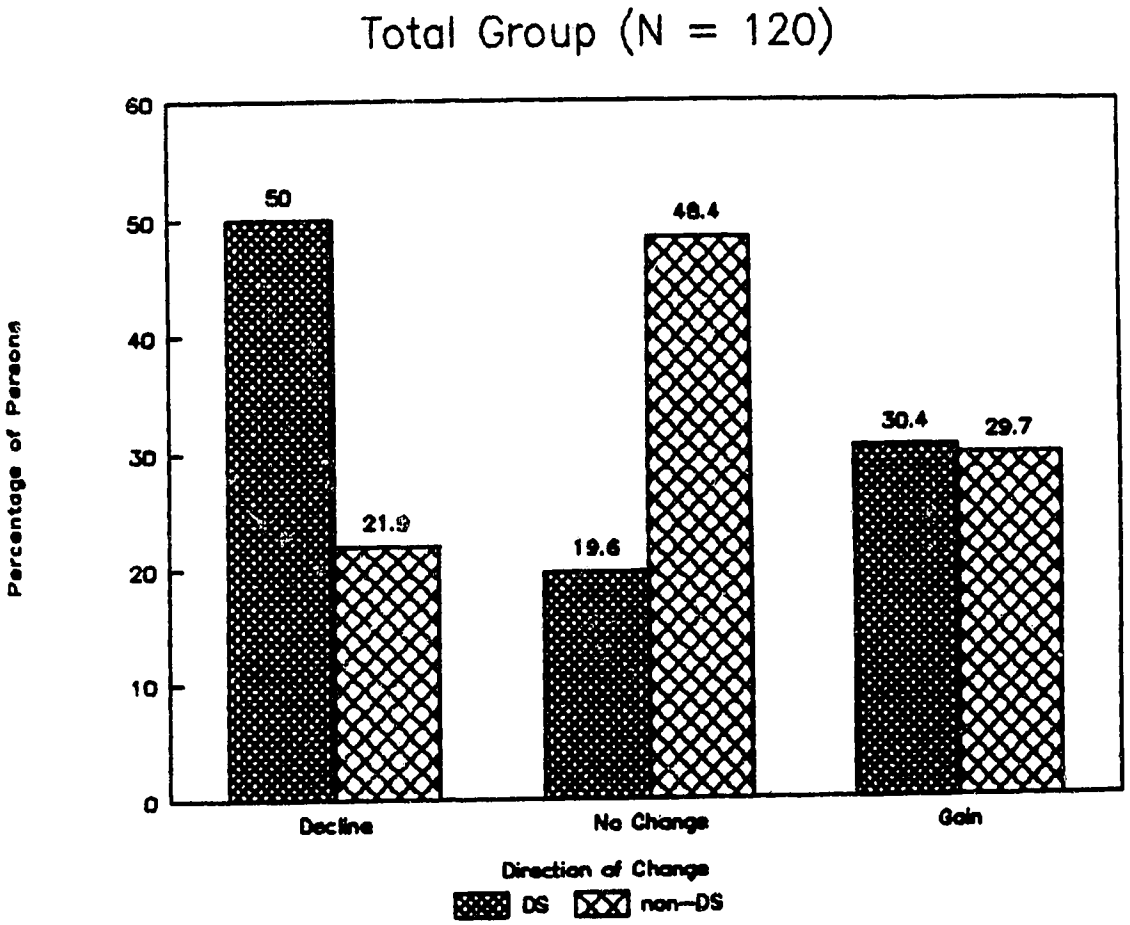
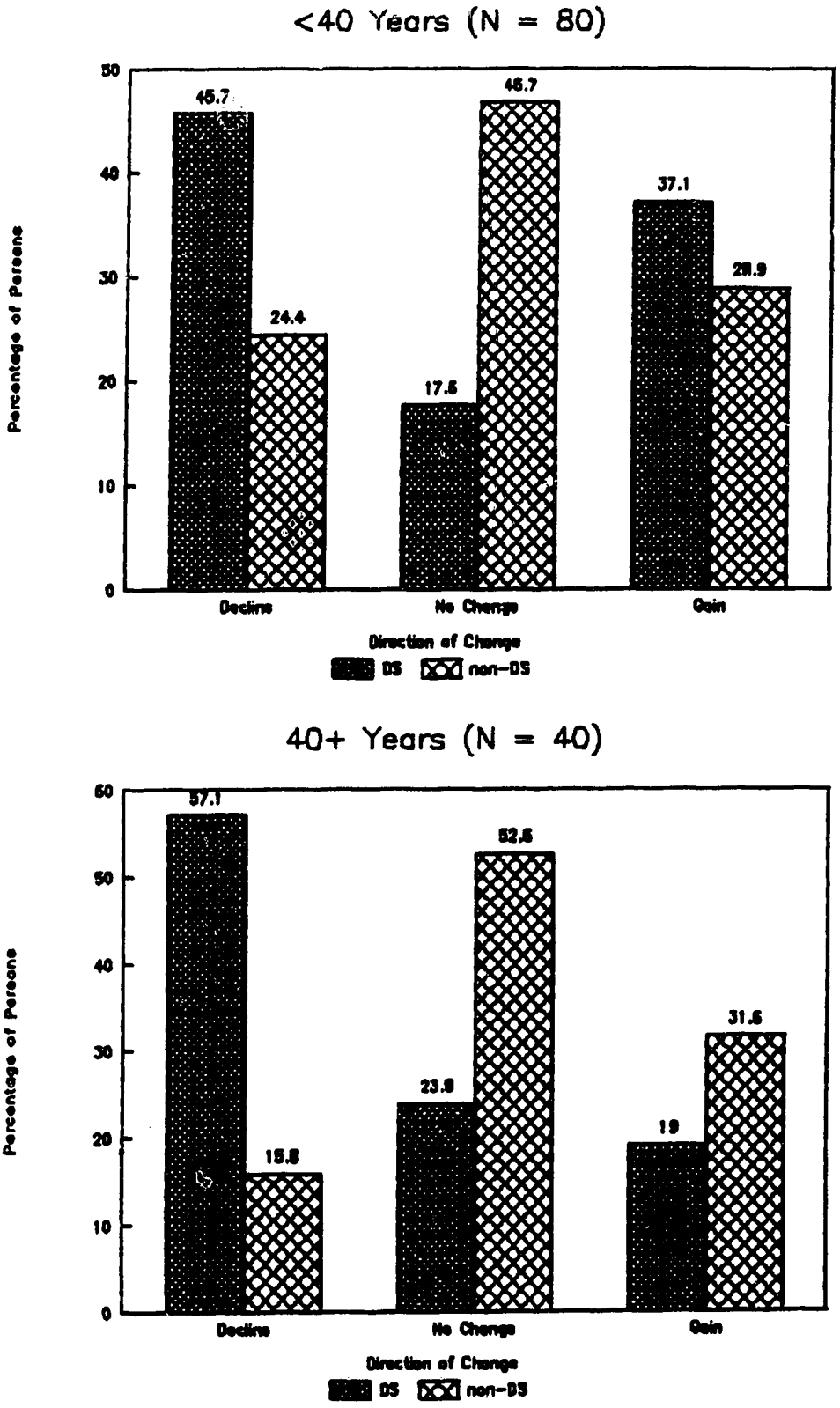


Figure 9 Continued



Numbers [$X^2(2) = 7.86$, $p<.019$] for the under 40 age group; and Domestic Behavior [$X^2(2) = 12.26$, $p<.002$], and Washing/Grooming [$X^2(2) = 6.48$, $p<.039$] for the over 40 age group. In Eating, Dressing, Domestic Behavior, and Washing/Grooming, the difference was attributed to a greater number of the DS group than expected recording declines (34.3%, 48.6%, 52.4%, and 57.1% respectively). In the Numbers subdomain a greater number of DS persons (77.1%) recorded no change in score while more non-DS persons than expected (28.9%) recorded gains.

By individual. Results indicated that 39% of DS individuals (22 of 56 persons), and 14% of non-DS adults (9 of 64 persons) recorded clinically significant declines in their general level of adaptive behavior (i.e., on the mean of 20 subdomains) on retest. Individuals were identified, and test reports from pretest and retest periods were reviewed. Explanations for decline in 7 of the 9 non-DS persons targeted could not be pinpointed from caregiver descriptions. In one case a change in residence with resultant uncooperative behavior, and in another, a history of epilepsy were suggested. Of the 22 DS individuals who recorded mean score losses, noticeable declines were mentioned in sensory-motor skills for 9 persons; in memory, cognitive skills, expressive language, and self-help skills for 2 individuals; and per a history of life-long severe self-abuse in one woman. For 8 DS individuals, no

explanation was indicated in test reports; while in 2 persons, transfer to a residence for higher functioning adults with resultant passivity, or assessment by an external psychologist unfamiliar with the DS population were suggested as possible reasons for mean score decline.

Subsequently, for each individual, a hierarchy of adaptive skill decline was determined through comparison of percentage competency per subdomain at pretest and retest. Only subdomains with declines in competency of 30% or greater were targeted initially. Changes for the 7 DS and 4 non-DS persons identified via this criteria are presented in Table 5. A similar pattern of decline across persons with severe/profound retardation included decrements in Self-Help and Communication domains as reported previously, with specific declines in washing/grooming, dressing, receptive language, and social interaction skills. The above findings, in conjunction with reports from current research suggest that 5 persons with DS were highly suspect for Alzheimer-like changes. In the 2 younger DS persons, blindness was suggested as reason for subdomain decline. Of the remaining DS persons who recorded less than a 30% decline per subdomain: 4 presented with primary vision and gross motor handicaps; 2, aged 44 and 51 years, and 4 less than 40 years of age recorded similar decline patterns of lesser magnitude and were therefore suspect for early

Table 5 Hierarchy of Skill Loss for Selected Individuals with Clinically Significant Declines in Mean Score at Retest

Etiology	Age	F/L	Percentage Decline per Subdomain
Greater than 30% Decline			
DS	66	s/p	TOIL (-60), GMOT (-40), RLAN (-38), WASH (-32).
DS	59	s/p	WASH (-42), DRES (-28), RLAN (-28).
DS	58	s/p	DRES (-62), WASH (-61), RLAN (-41), TOIL (-41), WRIT (-38), FMOT (-28), TR (-28).
DS	52	s/p	WASH (-38), RLAN (-38).
DS	48	m/m	DOME (-34), TIME (-34), SOCI (-31).
DS *	30	s/p	TR (-32), EAT (-31).
DS *	25	s/p	AR (-56), SOCI (-35), TR (-31), VOC (-31), REC (-30).
non-DS **	53	m/m	WRIT (-34).
non-DS	34	s/p	ELAN (-69), WASH (-47).
non-DS	33	s/p	WASH (-32).
non-DS	31	s/p	RLAN (-35), TR (-34).
Less than 30% Decline			
DS	51	s/p	SOCI (-28), WASH (-18), DOME (-16), EAT (-13), TR (-12), DRES (-10).
DS	44	s/p	VR (-22), WASH (-15), DOME (-13), RLAN (-13), TOIL (-12), SOCI (-10).
DS	33	s/p	WASH (-29), FMOT (-25), WRIT (-22), ELAN (-22), DRES (-19), SOCI (-19).
DS	33	s/p	RLAN (-32), ELAN (-22), DRES (-19), SOCI (-15).
DS	29	s/p	WASH (-29), TR (-22), EAT (-15), TOIL (-12), FMOT (-10), SOCI (-10).
DS	27	s/p	DRES (-28), RLAN (-25), TOIL (-22).

* Blind ** Epilepsy

Alzheimer-like changes. Hierarchies for these individuals are also presented in Table 5.

In the non-DS group, only 1 person over 40 years of age recorded a clinically significant decline in general level of adaptive behavior, possibly attributed to severe epilepsy. Of the under 40 year old group, a consistent hierarchy of skill decline was not evident.

Case studies. Similar hierarchies of skill loss in DS individuals recording gains in general level of adaptive behavior at retest indicated that the pattern of decline, rather than general level of adaptive behavior (i.e., mean score), was a more likely predictor of possible Alzheimer-like changes. This was demonstrated by tracing the progression of decline across 3 DS persons who died prior to data analysis for whom retest data and neuropathological findings were available. A fourth DS adult excluded by oversight from the study, but who recorded extensive neuropathology was also tracked.

Case #1 the oldest DS subject retested, a 69 year old blind male who had resided with his family until age 45, was reported to be a friendly, cooperative, obedient person who enjoyed social activities and particularly liked to be with children. Noticeable changes in fine motor skills and episodes of antisocial behavior began at age 62. Three weeks before death he recorded 8% in general level of adaptive behavior, down from 11% at pretest. Although

cognitive skills, receptive language, and domestic behavior were already at a floor, the hierarchy of skill loss for this person matched those above. Table 6 tracks the timing and sequence of skill decline recorded from caregiver descriptions compiled for annual Individual Program Plan (IPP) conferences held in a cumulative file. Cause of death was cardiac arrest and pneumonia. Extensive Alzheimer-type degeneration of the nervous system was found on autopsy (Clark, 1990).

Cases #2 and #3 were DS adults, both 54 years of age at retest each of whom recorded a 7% increase in general level of adaptive behavior. Case #2, a male institutionalized since age 36, tracked in Table 7 lost dressing and hygiene skills progressively, but maintained eating, toileting, and motor skills until death. It is noteworthy that social skills were maintained throughout the progression of decline in this individual, with a craving for attention, hugs, and hand-holding noted by caregivers 2 years prior to death. Family contact had virtually ceased upon admission to care. Assessment of adaptive behavior 4.5 years prior to death by a psychologist unfamiliar with the mentally handicapped population may reflect measurement error, and may have contributed to initial low scores. Subsequent re-assessment 1.5 years prior to death likely gave a truer estimate of adaptive behavior. Death resulted from pneumonia and cardiorespiratory arrest following an initial

Table 6 Case Study #1 - Progression of Decline in Adaptive Function

Prior to Death	Noted By	Behavior
11 years (age 58)	Psy	- has all functional skills - very social, friendly, cooperative - enjoys music, sings, dances, claps to music.
7 years	Res	* - fine motor skills decrease (drops objects) - walks independently * - withdrawn, rebellious behavior, strips clothing.
5 years	Psy	* - totally dependent in grooming * - dressing skills decline (can't tie or button) - likes to sing, dance, outings.
3 years	Med Psy	* - seizures begin (the only 2 seizures reported) * - totally dependent in dressing, washing, toileting, social interaction * - eats hand-over-hand * - ambulatory but must be led * - fine motor poor (unable to hold objects) * - no functional receptive/expressive language * - very passive, sleeps alot - recommendation: maintain gross motor skills.
1 year	Res	* - bedridden * - tactile responsiveness poor - no response to texture, does respond to cool * - no fine motor, or receptive language skills.
3 weeks (age 69)	Psy	* - very little tactile, or auditory responsiveness.
Autopsy		- severe loss of pyramidal cells in hippocampus, with neurofibrillary tangles and granulovacuolar degeneration in residual cells - numerous neurofibrillary tangles and plaques of the Alzheimer-type (59 tangles per mm^2, 118 plaques per mm^2) - extensive Alzheimer-type degeneration of the nervous system (Clark, 1990).

Note: Psy = Psychology Res = Residential Med = Medical

* Change from previous function first noted

Table 7 Case Study #2 - Progression of Decline in Adaptive Function

Prior to Death	Noted By	Behavior
8 years (age 46)	Psy	- has all functional skills - independent in dressing.
5 years	Psy	* - recommendation: encourage social & leisure skills
4 years	Res	* - recommendation: increase self-help skills (dressing, hygiene).
3.5 years	Psy	- very social - independent in eating, toileting, undressing - gross motor skills good (climbs stairs) - fine motor skills good (colors, uses pencil) * - dressing - needs assistance to pull up pants * - needs assistance to wash hands & face.
1.5 years	Psy	- tactile & auditory responsiveness adequate - social interaction - recognizes staff - gross motor skills good (climbs stairs) - fine motor skills good (uses pencil) - dressing - needs help to pull up pants * - totally dependent in washing - recommendation: increase self-help & gross motor skills.
3 months (age 54)	Res	- independent in eating & toileting - needs assistance in dressing - very good gross motor & fine motor skills (runs & jumps often) * - requires total care in washing/hygiene * - limited communication & understanding (doesn't respond to name) - behavior - cooperative & well behaved, cheerful & even tempered, but strips clothing.

Note: Psy = Psychology Res = Residential

* Change from previous function first noted

slow progression of suspected Alzheimer-like changes, then dramatic decline to death.

Case #3 documented in Table 8, a female institutionalized since age 23, recorded initial declines in domestic skills followed by withdrawal, disorientation, and difficulty with skills using fine motor sequences. Interestingly, early decline in function was also noted in the vocational setting where initial high performance began to slip. Sociability and memory for complex tasks began to decline 3 years prior to death. Ability to recognize caregivers became inconsistent and resulted in mood swings and fear responses. Levels of adaptive behavior assessed 4 years, and then 14 months before death indicated that most functional skills were present. An abrupt decline in all functional behavior 8 months before death, in actuality a dramatic 3 week period of skill loss, signaled onset of typical late Alzheimer-like changes. Pneumonia with respiratory failure was the reported cause of death. Extensive Alzheimer-type degeneration of the nervous system was reported at autopsy (Clark, 1990).

Case #4, a male institutionalized for medical reasons since age 6, presented an especially remarkable history. This individual tracked in Table 9 displayed an unusual sequence of behavior change. Early apprehension at leaving the residence, and deficits in memory, dressing, use of fasteners, and hygiene, all typical Alzheimer-like changes,

Table 8 Case Study #3 - Progression of Decline in Adaptive Function

Prior to Death	Noted By	Behavior
9 years (age 45)	Psy	- has all functional skills - independent in self-help skills - happy, social person, cooperative, well-behaved - recommendation: teach domestic skills (vacuuming, dusting, table setting).
6 years	Voc	- works independently, tabulates own production.
5 years	Res	* - inappropriate dress for weather.
4 years	Psy	- good self-help & receptive/expressive language skills - good fine motor skills (uses pencil) * - moderate skills in vocational, recreation, & domestic area (able to set table & dust)
	Res	- happy, pleasant, self-confident individual - inappropriate dress for weather.
3 years	Res	- all skills fully functional - helps staff do dishes & fold towels - inappropriate dress for weather * - prefers to be alone, refuses group activities * - doesn't want off-unit activities, becomes verbally abusive & cries when taken off-unit.
2.5 years	Res	* - unable to find way from vocational to residence * - disoriented on-unit * - forgets how to do simple tasks (ie., cleaning bathroom).
2 years	Res	- excellent functional skills - independent in eating (pours, cuts, uses napkin), toileting, dressing, & most washing/grooming (washes hair) skills * - needs verbal prompts for toothbrushing, clothes selection, changing clothes - good language skills - good gross motor/fine motor skills (climbs stairs, uses scissors) - loner * - seizures begin - recommendation: maintain socialization & self help skills (toileting, dressing, washing).

Table 8 Case Study #3 Continued

Prior to Death	Noted By	Behavior
14 months	Med	- epilepsy, hypothyroidism
	Res	- gross motor good (walks, runs, kicks, stairs)
		- independent in toileting, dressing (can button, zip), particular about what to wear
		* - inappropriate dress for weather
		* - eats with spoon - difficulty with utensils
		- uses pencil
		- bathes with assistance, attempts to wash hair
		- domestic behavior good (makes bed, tidies drawers, folds towels)
		- likes music, dancing
		- well behaved, happy, cooperative
	Psy	* - recommend: improve self-care & domestic skills.
8 months	OT	* - abrupt change in function
		* - fed self until 10 months before death
		* - neurologic changes noted (12 months to death).
6 months	Res	* - dramatic decline in overall function
		* - transferred to medical area.
2 months (age 54)	Med	- seizures diagnosed 2 years prior
		- dementia - (mild) began 2 years prior, Alzheimer's disease diagnosed 1 year prior
		- hypothyroidism diagnosed 2 years prior
	Res	* - tactile responsiveness poor
		* - gross motor poor (needs help to sit, stand, walk)
		* - fine motor poor (will not grasp, can't manipulate utensils)
		* - totally dependent in toileting, dressing, washing
		* - very passive
		- expressive language poor (uses one word if asked)
Autopsy	- almost total loss of pyramidal cells in hippocampus, conspicuous gliosis of neocortex	
	- neurofibrillary tangles (88 per mm^2), plaques (119 per mm^2) in parietal cortex	
	- extensive Alzheimer-type degeneration in brain (Clark, 1990).	

Note: Psy = Psychology Voc = Vocational Res = Residential
Med = Medical OT = Occupational Therapy

* Change from previous function first noted

Table 9 Case Study #4 - Progression of Decline in Adaptive Function

Prior to Death	Noted By	Behavior
9 years (age 36)	Res	- has all functional skills - hygiene good, makes bed, ties shoes, utensils - gross & fine motor good, swims (front & back crawl), musical instrument, claps in rhythm, does puzzles with color & shape discrimination - understands, performs 2 sequence instructions - polite, well behaved, never upset - works independently on verbal & visual directions
8 years	Res	- good table habits (utensils, pours, serves) & hygiene (bathes, washes hair) - manipulates fasteners, scissors, draws * - doesn't want to leave residence alone, sits on lawn & hallucinates * - cognitive skills "can't remember a thing" (time, dates, weeks, months) - well behaved, follows instructions
	Voc	* - no concept of time * - poor hygiene (messy dress & hygiene).
7 years	Med	- difficulty chewing food (chokes at times) - hearing adequate, refuses to come at times
	Res	- eating skills good * - reminders to zip, button, brush hair & teeth, wash, wear socks & underclothes - attends social functions, no behavior problems - recommend: train table setting, laundry, room cleaning, dress, hygiene
	Voc	- clothing seldom clean & tidy, neglects buttons, belt, tucking shirt.
6.5 years	Voc	* - placement terminated for lack of progress * - tasks mixed up, can't apply words used at work, difficulty following instructions, needs direct supervision & prompts, seldom interacts - clothing dishevelled.
6 years	Res	- good mealtime skills, agile, cuts, pours, pencil - independent on grounds, makes decisions to attend social events, waits turn, shares.
5 years	Res	* - needs assistance with all self-help skills * - severe self-abuse (Pre-voc terminated).

Table 9 Case Study #4 Continued

Prior to Death	Noted By		Behavior
4 years	Res		- hallucinations, no self-abuse
			- recommend: teach self serve meals, bathing, non -escort. Clothes choosing training successful.
3 years	Res		- unescorted on grounds.
2 years	Psy	*	- needs repetition of stimulus words during testing
			- self-help skills relatively well developed
			- recommend: teach domestic skills (cleaning after self, care of clothing)
	Res		- independent in self-serve meals, uses utensils
			- independent in self-help skills, selects & puts clothing away, supervision for buttoning, shaving
			- unescorted on grounds, enjoys community & overnight outings
			- recommend: teach buttoning
	Voc	*	- inappropriate grooming & hygiene, doesn't dress for weather.
1 year	Med		- collapsed twice (seizures?)
	Res		- successful completion of buttoning program
			- mealtime skills excellent, independent in toileting, bathing, hair washing (needs help to comb & shave)
			- chooses clothing, zips, buttons, ties shoes, assistance for clothing for weather
		*	- loner, needs supervision off-unit (wanders, stares)
		*	- often confused, needs time to understand
			- cooperative, well behaved.
9 months	PreVoc	*	- inappropriate grooming, hygiene major problem
			- placement terminated for lack of progress
		*	- tries hard but frustrated & cries
	Med		- unable to find reason for regression
	Res	*	- can't do tasks, confused, frustrated.
1 month (age 45)	Med	*	- seizures infrequent, unsteady, collapses, falls
			- chokes easily
	Psy		- good gross & fine motor skills
		*	- no recognition, awareness, interest, understanding
			- extreme decline in adaptive behavior, no maladaptive behavior

Table 9 Case Study #4 Continued

Prior to Death	Noted By	Behavior
	Res	* - finger feeds unless prompted, spoon upside-down, plate guard, to shaky to pour
		* - forgets how to dress & undress, stares at clothing, waits for assistance, incontinent, can't find bathroom, forgets what to do
		- no awareness or understanding
		* - unsteady, falls, loses grasp (drops objects)
		* - very disoriented, loses thoughts during task, wanders.
Autopsy		- chronic CNS degeneration of Alzheimer type
		- enormous numbers of plaques (68 per mm^2), & tangles (73 per mm^2) in neocortex & hippocampus
		- extensive Alzheimer-type degeneration in brain (Clark, 1988).

Note: Res = Residential Voc = Vocational Med = Medical
Psy = Psychology PreVoc = Prevocational.

* Change from previous function first noted

were ameliorated by training. Caregivers targeted declining skills and successfully returned the individual to previous functioning levels. Initial decline of function was most evident in the vocational setting where poor time concept, poor hygiene, and messy dress preceeded difficulty with following instructions and completing tasks. Vocational and pre-vocational placements were terminated as a result. Eventually confusion and wandering behavior preceeded rapid decline in all skills a month before death. Neuropathological evidence indicated that this individual had extensive Alzheimer type degeneration. Large counts of neurofibrillary tangles and neuritic plaques in the neocortex and hippocampal regions of the brain were reported (Clark, 1988). The neuropathologist verbally reported that decrements in mRNA usually reported in AD in the general population were absent in this case. Rather, this individual maintained mRNA levels expected of a 15 year old DS person with no AD changes. Death resulted from pneumonia.

The case studies illustrate a pattern of adaptive behavior change in aging DS centering on self-help and communication skill declines. This pattern was reported regardless of gains or losses on the general level of adaptive behavior at retest and seemed to begin with changes in neatness of dress, and the ability to manipulate fasteners. Interestingly, this was the subdomain on which a

clinically significant decline was reported by the under 40 year old DS group, and may signal early Alzheimer-like changes. Conversely, both DS persons beyond 65 years of age recorded the most pronounced change in toileting and gross motor skills, symptoms of late AD in the general population. Preliminary evidence gained from tracking caregiver descriptions of 4 deceased DS persons indicated that the onset and rate of progression tended to vary with the individual, although the terminal stage in 3 of 4 persons mimicked late AD in the general population. Dramatic acceleration of the disease process prior to death was indicated in two 54 year old persons. Alzheimer's disease was confirmed in all 4 individuals at post-mortem.

Summary

Results received throughout this study were generally consistent. Cross-sectional examination of distinct groups revealed that differences in adaptive behavior were related to functioning level rather than to etiology or to age. Down syndrome adults showed significantly greater competence in Self-Help and Socialization domains than their non-Down syndrome peers, however. Age-related differences in adaptive behavior were not reported via this methodology.

Longitudinal examination of the same individuals following a 3 to 4 year retest interval demonstrated consistency in profiles over time, in both DS and non-DS groups. A trend emerged for DS persons beyond age 40

however, where a greater number of persons recorded less stability in their general level of adaptive behavior with age, with significant numbers of people declining in Self-Help and Communication domains. This finding was supported via comparison of pretest and retest means, where clinically significant decreases in adaptive behavior occurred in the over 40 year old DS group in washing/grooming, gross motor, receptive language, social interaction, dressing, domestic behavior, and toileting skills. Similar patterns of subdomain decline in 5 DS persons beyond 48 years of age were highly suspect for Alzheimer-like changes. The progression of adaptive skill decline in 4 DS persons with confirmed AD at post-mortem indicated that the pattern of skill decline was a more important predictor of possible AD in DS than were changes in the general level of adaptive behavior.

CHAPTER V

Discussion

This study examined the effects of age on adaptive behavior of mentally handicapped adults. Results will be discussed in relation to methodologies used, the aging process in DS, and the likelihood of aging DS persons displaying AD.

Research Design

Two methodological approaches employed in this study have produced seemingly contradictory answers to the research question investigated. Each design examined a different aspect of the problem however.

Cross-sectional methodology revealed that no age-related differences existed between DS and non-DS groups. By comparing distinct groups of individuals differing in etiology, age, and functioning level, at one particular point in their life, it was not possible to determine if changes with age had occurred. Instead results indicated that DS groups do display greater competence than agemates in all adaptive skill domains excluding the communication domain where hearing deficits accompany the condition, and that they show particular ability in self-help skills and social skills. The conclusion drawn indicates that adaptive behavior does not differ between groups of different ages.

Several factors may contribute to these findings. Evidence for a premature aging hypothesis (Cunningham, 1982; Gibson, 1978; St Clair & Blackwood, 1985), and mortality curves advanced by 20 years in aging DS persons (Sinex, 1986; Thase, 1982) suggest that in older DS groups death has already claimed individuals who are less healthy or who have succumbed to old age, leaving behind a group not equivalent to their non-DS cohorts. A higher skill level in the aged DS group might be a related outcome.

Institutional placement for individuals has been considered when behavioral, medical, or family issues could not be resolved through less intrusive measures. Therefore, younger admissions to care tend to have more severe skill deficits or behavior problems and tend to remain longer in the institutional environment. Older adults, conversely, are often admitted when aging family members can no longer provide care in the home. Years of community living contribute to greater competence in self-care and social skills in these individuals. It is noteworthy that in this study individuals had been institutionalized for an average of 19 years in the under 40 group (age range 18 to 39 years), and 27.7 years in the over 40 group (age range 40 to 75 years). This would indicate that younger groups have spent a greater proportion of their lifespan in an institutional setting than have older age groups. Levels of skill competence may be a reflection of this.

Finally, the effects of training may contribute to maintenance of skill levels in older groups. Mentally handicapped persons typically require more intensive teaching methods, over a longer period of time, to acquire lasting behavior change. Although both age groups, regardless of etiology, have had the benefit of training, the older group has also had the benefit of time. Skill maintenance is a likely outcome. The above explanations offer support for the stability of adaptive behavior in older age groups regardless of etiology.

In this study, cross-sectional design had its limitations. Longitudinal methodology was better able to address the research question: Do adaptive skills in DS adults show a greater decline with age than adaptive skills of non-DS mentally handicapped adults? The remainder of this chapter will focus on findings gained via this methodology.

Evidence for Alzheimer's Disease in Down Syndrome

Results of the present study indicate that adaptive skills in DS adults are more likely to decline with age than that reported by mentally handicapped adults of other etiologies. Not only did the general level of adaptive behavior show less stability for a greater number of DS persons beyond age 40, but a pattern of skill decline was evident with age. Findings suggest that for some persons early decrements begin with dressing and vocational skills

in younger DS persons, and that as they surpass 40 years of age, self-help skills including hygiene, dressing, and toileting, receptive language, social interaction, domestic behavior, and for some persons gross motor skills, record declines to the extent that additional prompts and assistance are required in daily routines. Stability or gain in adaptive behavior at retest in non-DS persons indicate that the pattern shown in DS is unlikely a response to normal aging.

Previously, studies investigated the discrepancy between neuropathological and clinical evidence for AD in aging DS persons. So far the evidence accumulated has found an exact parallel between AD and the effect of aging in DS on an anatomical and biochemical level. It is at the behavioral level that a different perspective emerges and it has been difficult to reconcile clinical observations with laboratory findings. Some researchers have suggested that what develops in DS adults is not AD but some form of a mimicking condition. Some believe that all older DS persons have AD on anatomical grounds (Mann, 1988). Some search for subtle changes in behavior in aging DS persons (Lai & Williams, 1989). Studies have primarily targeted cognitive skill changes and secondarily, adaptive skill changes as indicators of the disease presence and progression in an attempt to delineate the prevalence, age at onset, and course of AD in DS.

Course of the Disease

Several studies have reported that the course of AD in those DS individuals who display dementia, differs from that found in the general population. Schweber (1988) emphasized the difficulty of diagnosing dementia in the DS population, and noted that in institutional records a diagnosis is now applied after an individual undergoes marked personality change, develops incontinence, becomes hostile, apathetic, and withdrawn, and frequently develops late onset seizures. These symptoms, however, are characteristic of the very last stages of AD in the non-retarded population, and do little to make understandable clinical versus laboratory findings. The most reasonable conclusion is that AD truly is expressed differently in those with or without DS. Where anatomical, biochemical, and behavioral changes are manifested in AD in the general population, only anatomical and biochemical alterations typify the disease in DS. Schweber (1988) proposed that classification in DS might be (1) "quiescent" AD if no gross behavioral changes occurred, (2) "partial" AD if seizures without dementia occurred, or (3) "active" AD if both seizures and dementia were displayed.

Other investigators reporting decline in self-care skills align yet more closely with the findings in the present study. Miniszek (1983) suggested that older DS persons with or without dementia were differentiated by their profile of adaptive behavior. Extremely uniform

patterns were typical of the regressed group, and greater diversity in profiles were typical of the non-regressed group. Zigman, Schupf, Lubin, and Silverman (1987) similarly recorded decline in adaptive skills in aging DS persons and found the pattern of age-related deficits were not affected by developmental level nor by community versus institutional placement. Finally, Lai and Williams (1989) delineated the 3 stages of clinical deterioration for DS persons with AD as: (1) temporal and spatial disorientation, decreased social interaction and inattention; (2) loss of self-help skills such as dressing, toileting, and use of food utensils, gait disorders, and often seizures; and (3) incontinence in non-ambulatory bedridden patients often accompanying pathological reflexes which preceded death from pneumonia and infection.

Although the present study was able to delineate a pattern of skill decline, the author can only speculate as to the presence and progression of AD in subjects still living. It does appear that a number of older DS subjects are involved in a pattern of skill loss akin to those followed by Lai and Williams prior to death. It does appear that more than "quiescent" AD changes have occurred in several persons beyond 40 years of age, although 4 individuals aged 27 to 33 years also recorded suspect profile changes of lesser magnitude. It does appear that DS persons beyond 50 years record losses of yet greater

magnitude, and that the 2 individuals beyond 60 years showed their greatest declines in toileting and gross motor skills.

Only in those individuals, who died prior to data analysis and for whom neuropathological data were available, can conclusive statements as to the presence of AD be made. Although these persons did not record clinically significant declines in their general level of adaptive behavior on retest, due to the timing of the assessment cycle, and to their rapid decline, they did demonstrate the pattern of skill loss isolated in this study, and were observed by caregivers in the years preceding death to show "typical" DS Alzheimer-like changes in personality, orientation, and self-help skills. According to Schweber's (1988) classification, "active" AD was present in cases #1, #3, and #4.

Important conclusions can be drawn from these 4 subjects. First, the pattern of skill decline is a more likely predictor of AD in DS than is a decrease in the general level of adaptive behavior. Secondly, progression of the disease is variable - some individuals proceed slowly through skill loss, others demonstrate a dramatic decrease in many adaptive skills just prior to death. Thirdly, case #4 offers hope for active treatment of AD in the preliminary stages. Successful training programs alleviated skill decrements in clothing selection, buttoning, and unescorted mobility in the early stages of the illness and facilitated

independence for a longer period. Finally, although an adaptive assessment tool structures behavioral observation and reporting, alert caregivers offer the greatest hope for early detection of possible Alzheimer-like changes in the DS population. Identification of early indicators in the disease process will assist caregivers in their task. Results of this study suggest that the need for help with hygiene and fasteners used in dressing may serve as the first clues to decline. Although Lott and Lai (1982) also found this as the first evidence of decline, they felt that it was the motivation that was lost before the actual ability to perform these activities. This may well be an accurate assumption, since a strength of behavior management procedures which are frequently used with low functioning populations is to boost motivation to perform skills that are already in the individual's repertoire.

Prevalence

An estimate of the prevalence of AD in the DS population would enable appropriate services to be established for those facing terminal skill loss. In the present study the incidence of Alzheimer-like involvement is difficult to guage. Given that the pattern of skill loss was found in individuals recording both decline or stability of general level of adaptive behavior at retest, our estimate of possible Alzheimer-type involvement in 39% of the DS sample seems a conservative albeit speculative one.

Lai and Williams (1989) found the prevalence of dementia in the institutionalized DS population to be 8% between 35 to 49 years, 55% between 50 to 59 years, and 75% of those over 60 years, generally paralleling the increased incidence and severity with age found in the present study.

In the general population, prevalence rates were found to be a function of the assessment instruments used, and their sensitivity in detecting various levels of dementia (Black, Blessed, Edwardson, & Kay, 1990; Copeland, 1990). Diagnosis of dementia in these studies relied heavily on clinical judgements of psychiatrists. Black and colleagues (1990) stressed that when comparing studies, it was important to know the level of dementia that the instrument could detect. Use of adaptive skills as predictors of dementia is not without problems either however. Individual variability in both symptoms and rate of progression underscore the need for caution in assuming a tentative diagnosis. In the final analysis and with the research currently available, diagnosis of AD for many DS individuals can only be made postmortem.

The atypical course of AD reported in DS further confounds estimates of disease incidence in this population. Early cognitive changes of AD are seldom reported in DS and debate continues as to whether this is a function of altered disease progression, of inability to assess subtle changes in predominantly low functioning institutional samples

(Miniszek, 1983), or of dementia superimposed on a developmentally abnormal brain (Lai & Williams, 1989). In the present study, early cognitive changes typical of AD were absent, due likely to a predominance of subjects with severe/profound retardation. Cognitive skills were already at a floor in these individuals. Evidence for late Alzheimer-type changes were found through evaluation of skill decline with age, however. Perhaps what is needed is as Zigman and associates (1987) suggest - to revise DSM III criteria for the diagnosis of dementia to reflect its course in the DS population. A consistent marker, based on relevant research, would enable more accurate prevalence estimates to be made, and hence more suitable services to be established.

Age at Onset

Service provision would also benefit from knowledge of estimates of age at onset of the disease. Dalton and Crapper (1984) reported a range of 42 to 60 years at onset, and Wisniewski, Dalton, Crapper-McLachlan, Wen, and Wisniewski (1985), a range of 43 to 57 years. Zigman and colleagues (1987) found that average age at onset of dementia and/or behavioral regression was about 50 years of age. Lai and Williams (1989) similarly reported average age of onset to be 54.2 $\pm$ 6.1 years. In 4 case studies presented in this study, average age of onset of behavioral changes reported by caregivers was 49.2 years (37 years for

one person, 49 years for two, and 62 years for the other), demonstrating compliance with previous research. It appears that neuropathological changes found in the brains of DS persons beyond 35 years of age do not translate to behavioral changes for several years, if at all. Lai and Williams (1989) suggest such an incubation period could be as much as 20 to 30 years and could therefore explain why some DS persons with neuropathology lacked signs of clinical dementia at death.

Findings of the present study suggest that the presence of a threshold number of plaques and tangles, in critical regions of the brain (i.e., the hippocampus, and/or parietal cortex), may be necessary before neuroanatomical and biochemical changes culminate in behavioral decline. The incredibly rapid loss of functional skills recorded by cases #3 and #4 support this idea. In both cases a 3 week period of dramatic skill loss occurred, for #3 10 months before death, and for #4 culminating in death. Human beings are biological organisms, with cognitive abilities, functioning in a social world. Adaptive behavior is the observable outcome of this complex interrelationship. If we are to understand the behavior we see, it is imperative that we acknowledge the integration of physiological, psychological, and social aspects of the human organism. Behavioral regression, therefore, may result from underlying

physiological alterations which do not permit the individual to function to previous levels.

Services for aging DS persons, therefore, should include provisions for an active training component for those who maintain functional behavior at age. Sensitivity to the pattern of skill loss indicative of AD, additional assistance, structured living environment, emotional support, and reasonable expectations become important to those who begin the disease process. It is important that a range of services be provided for the aging DS individual.

Aging in Down Syndrome

Whether preceding deficits should be regarded as evidence of the early development of Alzheimer-type dementia in DS is however, still a subject of debate. Some investigators have ascribed such symptoms to other factors.

It was suggested previously that the developmental process appears to be qualitatively different in DS. A post-natal curtailment of growth and maturation in brain weight, head circumference, and development of individual neurons; and decreases in body weight and length during early childhood have been reported (Kemper,1988). Similarly, differences in learning patterns during the early years have been found (Gibson, 1966; Wishart, 1988). Later in the lifespan, a premature aging hypothesis has been proposed to explain the untimely appearance of a variety of age-related changes and early death (Cunningham, 1982;

Gibson, 1978; Pueschel, 1988; Sinex, 1986; St Clair & Blackwood, 1985; Thase, 1982). Accelerated neurological aging has likewise been reported in the literature (Eisner, 1983). This evidence when taken collectively suggests that DS persons are not subject to the same developmental process as are others.

It is well known that the appearance of senile plaques and neurofibrillary tangles are associated with normal aging in the general population, and tend to become more numerous with age. The premature aging hypothesis uses this line of reasoning to account for the presence of significant quantities of plaques and tangles in aging DS brains. However, it is unable to explain why the incidence of dementia in DS persons over 60 years of age is 3 times that recorded in those 80 years or older in the general population. Further, it is noteworthy that there is no evidence that the presence of a few plaques and tangles are associated with mild age-related decline in intellectual function (Wisniewski & Rabe, 1986). The presence, number, and location of senile plaques and neurofibrillary tangles in DS parallels that recorded in AD signifying that in this case a pathological, rather than an aging, process is in progress. To extend the explanation yet further, perhaps AD is a heritable condition subject to a gene located on the 21st chromosome, whose penetrance is a function of aging

(Sinex & Myers, 1982). Premature aging would therefore tend to bring the condition on earlier in DS.

Another factor which merits consideration before a diagnosis of AD can be attributed to the pattern of skill decline recorded by DS persons, revolves around the typical features of the condition itself. DS persons are unique. Their extra chromosomal material is responsible for a variety of features not commonly found in other etiologies of mental retardation, nor in members of the non-retarded population. A high incidence of hearing loss, reported to occur in as many as 2/3 of the DS population, may be related to receptive language deficits, decreased attention, and a lessened desire to socialize with age. Poor muscle tone and skeletal abnormalities make gait disturbances appear more pronounced as age emphasizes the problem. Thyroid deficiencies are known to occur increasingly with age in DS persons, resulting in delayed growth and short stature in the early years, and apathy and lethargy later in life (Odell, 1988). An abnormal or defective immune system, the effects of which increase with age, contributes to the high incidence of leukemia and additional health problems beyond 40 years of age (Odell, 1988). When taken together the typical features of DS may be responsible for many behavioral changes in this population, that are not expected in others.

Summary

It is critical that normal features of the DS condition be ruled out before a diagnosis of AD is attributed to patterns of behavior and skill loss found in aging DS persons. Premature aging may advance conditions normally experienced by others later in the lifespan. Alzheimer's neuropathology found in virtually 100% of DS persons beyond 35 years of age, suggest that behavioral changes late in the lifespan of DS individuals result from anatomical and biochemical alterations of the disease. A pattern of skill decline has been reported in DS persons beyond 40 years of age in this study. This pattern was seen in 4 individuals who died from pneumonia following an Alzheimer's progression. However given the small sample size, it is still speculative as to whether these changes are universally indicative of Alzheimer's disease in Down syndrome.

CHAPTER VI

Conclusions

Findings of the present study suggest tentative conclusions regarding the effects of age on adaptive behavior in mentally handicapped persons. Age does not appear to unduly affect adaptive behavior in institutionalized mentally handicapped adults across a variety of etiologies. Stability of profiles in both younger and older adults was more the norm given a 3 to 4 year interval between assessments.

Individuals with Down syndrome however, and most particularly those beyond 40 years of age, appear to be at greater risk for decreases in adaptive behavior. Personal hygiene, neatness of dress, ability to comprehend language, and desire to socialize show decrements in many older DS adults. This may be related to the unusual developmental process that occurs throughout the lifespan, in concert with the typical features accompanying this unique chromosomal condition.

The incidence of dementia, three times that found in the general population, onset 20 years earlier, and extensive neuropathological changes in the brains of virtually all Down syndrome persons beyond 35 years of age, strongly suggest that behavioral changes reported in aging

persons with Down syndrome are those of Alzheimer's disease. Less than 100% compliance between that recorded on a molecular level, and that observed at a behavioral level suggest caution in attributing behavioral outcomes to biochemical causes. Rather, clinical decisions should take into account all sources of information. Individuals should be screened, and where necessary, treated for conditions such as hypothyroidism and hearing loss before an Alzheimer's diagnosis is entertained.

It would be inaccurate to consider neuropathological evidence in isolation from psychological status, and to assume that changes at a molecular level translate to clinical decline. Research does not show that to be true. At the same time it would be unfair to place unreasonable performance expectations on individuals involved in a progressive and terminal disease process. Understanding and support are required to help them cope with the confusion they encounter. Sensitive and alert caregivers hold the key to a happy and successful aging process for all individuals.

Implications for Service Providers

Several practical implications for service providers can be drawn from the present study. Of primary importance, is the need to assist all mentally handicapped persons develop to their fullest potential in behavioral, psychological, and social spheres. Given that DS adults have a greater risk for skill decline beyond 40 years of

age, it is even more critical that this unique group develop a solid base of functional behavior to enhance their adult years. Habilitation in areas of daily living skills, social situations, cognitive abilities, and personal relationships should be ongoing. DS persons and their mentally handicapped cohorts will always benefit from involvement and instruction. Service providers are entrusted with this responsibility.

The need for quality, routine, medical care in the DS population was highlighted by the findings of this study. Deficits in hearing and vision, thyroid deficiencies, and seizure activity, tend to become more predominant with age for this group. These conditions are amenable to medical intervention and help to enhance the well-being of the individual. It is particularly important to rule out these conditions before considering a less hopeful diagnosis of AD.

A routine adaptive behavior assessment regimen can be very helpful in charting individuals' abilities to cope with their environment. Skill changes may occur so gradually that they blend into the everyday milieu and go unnoticed by caregivers. A qualitative assessment of functional behavior can help to pinpoint behavioral decline, and signal the need for additional assistance or training. In this study, a 3 year interval between assessments missed critical behavioral

changes in some individuals. An annual assessment of adaptive behavior would be more helpful.

Finally, the findings in this study will help service providers to focus on key areas of behavioral decline as predictors of an AD progression. Caregivers should be alert to changes in the motivation to care for clothing, and in neatness of dress of their clients. Personality changes such as irritability, and fear of leaving familiar places also provide early clues. In higher functioning DS individuals, difficulty in coping with daily events, and in following instructions may be regarded as noncompliant behavior, but in fact signal early AD. The provision of additional prompts, assistance, and a more highly structured environment are more supportive and helpful interventions than introduction of a behavior management scheme to manage noncompliant behavior. A holistic approach to determining what is most helpful for the aging DS person will best serve their needs.

Limitations of the Study

The present study was subject to several methodological limitations which must be taken into account when evaluating the findings.

A retrospective study as was conducted relies completely on data recorded in the past. Only information deemed important at the time and noted down, was available for analysis. Incomplete description, assumptions as to

criteria used for assessing variables like deafness or time concepts, and inability to estimate the reliability among assessors were a reality. Interesting leads in the data had to be aborted if insufficient information was available in the records. By the same token, however, caregivers and assessors were not aware of the research to follow and therefore did not focus attention on the research question under investigation. This was a positive outcome of the retrospective nature of the study.

Generalization to the larger community of DS persons is facilitated when a representative sample constitutes the subject pool. In this study, the sample consisted of institutionalized individuals with an adaptive behavior assessment taken via the Pyramid Scales. Subject to subject matching on variables deemed relevant formed the criteria for inclusion in a comparison group. Given that diversity is the norm among mentally handicapped persons, that institutional populations tend to record greater deficiencies in adaptive behavior, and that a sample not representative of the global DS population was used, generality of findings is limited. Results may therefore be representative of predominantly low functioning DS persons residing in institutional settings.

Assessment of adaptive behavior is not without its problems. Reliability and validity of adaptive behavior assessment scales are difficult to determine. In addition,

scale items can mislead the assessor into believing systematic behavior change has occurred, when in fact ratio scales may display unequal interval characteristics. Recent development of The Pyramid Scales, its limited use in past research studies, and minimal information on its ability to detect change over time deprive us of the parameters necessary to effectively assess the stability of its results. Subjective interpretation of items by assessors may contribute to lack of consistency in assessment. All of the preceding factors may have influenced the reliability of results in the present study. It is wise to bear this in mind when evaluating the findings.

Reliance on a 3 to 4 year interval out of the space of a lifespan to provide indicators of adaptive behavior change, is perhaps an unreasonable window. Statements of long term change derive from long term follow-up. The results gained in this study should be considered in light of their short duration.

Finally, use of adaptive behavior measures to screen for dementia that by definition relates to cognitive changes, seems suspect. Given the population targeted in this study, and the exploratory state of current research, perhaps this limitation is more reasonably considered a strength. Investigators must begin with what subjects have to offer. Adaptive behavior competence determines how successfully individuals will cope with their world.

Indicators of the ability to cope must be readily available to caregivers. Adaptive behavior serves that function.

Future Research

The tendency, prevalence, and severity of adaptive behavior decline in aging Down syndrome individuals drives further investigation. Several recommendations for future research can be made.

Prospective studies geared to pinpoint the incidence, age at onset and significant behavioral indicators of adaptive skill decline in DS adults need to occur. Consistent long range tracking of behavior change will ultimately serve the individual best, and permit an accumulation of data from which generalizations may be drawn.

Research conducted previously by the medical community has focused on molecular changes per decade of life. Organization of behavioral data in the same manner may permit researchers to align findings and uncover the current mystery between discrepant laboratory and clinical evidence. Collaboration between disciplines can only advance knowledge in the area.

Assessment of dementia in the general population relies on verbally loaded screening devices to pinpoint change in cognitive functioning. Assessment of dementia in the mentally handicapped population requires devices sensitive to behavioral functioning. It is recommended that research

proceed toward the goal of identifying adaptive skills indicative of the disease, and toward subsequent development of a screening device to aid caregivers in assessment.

As the majority of Down syndrome individuals reside in community settings, representation from this sector in future studies is imperative. It is possible that a history of competence and independent living may provide a buffer to initial skill decline in those who would develop dementia. It is important to determine the extent to which disease progression is mediated by initial competence levels. Prevention or moderation of the disease process may be possible through ensuring Down syndrome individuals attain their full potential.

Finally, research to explore the parameters of intellectual and adaptive behavior within individual etiologies of mental retardation needs to occur. Trajectories of decline differ per etiology. Knowledge of within group differences would make judgements of change more realistic. A concept of normal variation per etiology would provide new standards in assessment and signal the need for appropriate intervention.

Mentally handicapped persons have long been regarded as second class citizens. Their unique problems and special needs merit our consideration.

REFERENCES

American Psychiatric Association. (1980). Diagnostic and statistical manual of mental disorders (3rd ed.). Washington DC: Author.

Barcikowska, M., Silverman, W., Zigman, W., Kozlowski, P., Kujawa, M., Rudelli, R., & Wisniewski, H. (1989). Alzheimer-type neuropathology and clinical symptoms of dementia in mentally retarded people without Down syndrome. American Journal on Mental Retardation, 93(5), 551-557.

Bauer, A., & Shea, T. (1986). Alzheimer's disease and Down's syndrome: A review and implications for adult services. Education and Training of the Mentally Retarded, 21(2), 144-150.

Becker, J., Huff, J., Nebes, R., Holland, A., & Boller, F. (1988). Neuropsychological function in Alzheimer's disease: Pattern of impairment and rates of progression. Archives of Neurology, 45, 263-268.

Berini, R., & Kahn, E. (1987). Clinical genetics handbook. New Jersey: National Genetics Foundation Inc.

Black, S., Blessed, G., Edwardson, J., & Kay, D. (1990). Prevalence rates of dementia in an aging population: Are low rates due to the use of insensitive instruments? Age and Aging, 19, 84-90.

Clark, A. (1988). Autopsy report. Calgary: Foothills Hospital.

Clark, A. (1990). Autopsy report. Calgary: Foothills Hospital.

Cone, J. (1984). The Pyramid Scales: Criterion referenced measures of adaptive behavior in severely handicapped persons. Austin: Pro.ed.

Copeland, J. (1990). Suitable instruments for detecting dementia in community samples. Age and Aging, 19, 81-83.

Cummings, J., & Benson, D. (1986). Dementia of the Alzheimer type: An inventory of diagnostic clinical features. Journal of the American Geriatrics Society, 34, 12-19.

Cunningham, C. (1982). Down's syndrome: An introduction for parents. London: The Trinity Press.

Dalton, A., & Crapper, D. (1984). Incidence of memory deterioration in ageing persons with Down's syndrome. In J. M. Berg (Ed.), Perspectives and progress in mental retardation (pp. 55-62). Baltimore: University Park Press.

Dalton, A., & Crapper-McLachlan, D. (1986). Clinical expression of Alzheimer's disease in Down's syndrome. Psychiatric Clinics of North America, 9, 659-670.

Dalton, A., Crapper, D., & Schlotterer, G. (1974). Alzheimer's disease in Down's syndrome: Visual retention deficits. Cortex, 10, 366-377.

Doyle, G., Dunn, S., Thadani, I., & Lenihan, P. (1986). Investigating tools to aid in restorative care for Alzheimer's patients. Journal of Gerontological Nursing, 12(9), 19-24.

Eisner, D. (1983). Down syndrome and aging: Is senile dementia inevitable? Psychological Reports, 52, 119-124.

Erickson, B., & Nosanchuk, T. (1979). Understanding data, Toronto: McGraw-Hill Ryerson Limited.

Fenner, M., Hewitt, K., & Torpy, D. (1987). Down's syndrome: Intellectual and behavioral functioning during adulthood. Journal of Mental Deficiency Research, 31, 241-249.

Fine, M., Tangeman, P., & Woodward, J. (1990). Changes in adaptive behavior of older adults with mental retardation following deinstitutionalization. American Journal on Mental Retardation, 94(6), 661-668.

Francis, S. (1970). Behavior of low-grade institutionalized mongoloids: Changes with age. American Journal of Mental Deficiency, 75(1), 92-101.

Gibson, D. (1966). Early developmental staging as a prophecy index in Down's syndrome. American Journal of Mental Deficiency, 70, 825-828.

Gibson, D. (1978). Down's syndrome: The psychology of mongolism. New York: Cambridge University Press.

Glenner, G. (1985). On causative theories in Alzheimer's disease. Human Pathology, 16(5), 433-435.

Grossman, H. (Ed.) (1983). Classification in mental retardation. Washington: American Association on Mental Deficiency.

Heston, L. (1984). Down syndrome and Alzheimer's dementia: Defining an association. Psychiatric Developments, 4, 287-294.

Hewitt, K., Carter, G., & Jancar, J. (1985). Ageing in Down's syndrome. British Journal of Psychiatry, 147, 58-62.

Hewitt, K., Fenner, M., & Torpy, D. (1986). Cognitive and behavioral profiles of the elderly mentally handicapped. Journal of Mental Deficiency Research, 30, 217-225.

Ho, J. (1989a). Psychological assessment at Michener Centre. Unpublished manuscript.

Ho, J. (1989b). Alzheimer's disease (AD) and Down's syndrome (DS): A preliminary study with 143 Michener Centre clients. Unpublished manuscript.

Hogg, J., & Raynes, N. (1987). Assessment in mental handicap: A guide to assessment practices, tests and checklists. London: Brookline Books.

Jervis, G. (1948). Early senile dementia in mongoloid idiocy. American Journal of Psychiatry, 105(2), 100-106.

Jolles, J. (1986). Cognitive, emotional and behavioral dysfunctions in aging and dementia. In D. Swaab, E. Fliers, M. Mirmiran, W. Van Gool, & F. Van Haaren (Eds.), Aging of the brain and Alzheimer's disease. Progress in Brain Research, 70, (pp. 15-39). New York: Elsevier.

Kaye, J. (1988). Neurochemical aspects of motor impairment. In R. Friedland (Mod.), Alzheimer disease: Clinical and biological heterogeneity. Annals of Internal Medicine, 109, 298-311.

Kemper, T. (1988). Neuropathology of Down syndrome. In L. Nadel (Ed.), The psychobiology of Down syndrome, (pp. 269-289). London: The MIT Press.

Kertesz, A., Appell, J., & Fisman, M. (1986). The dissolution of language in Alzheimer's disease. Canadian Journal of Neurological Sciences, 13, 415-418.

Khan, A. (1986). Clinical disorders of memory, New York: Plenum Medical Book Company.

Koss, E. (1988). Behavioral heterogeneity. In R. Friedland (Mod.), Alzheimer's disease: Clinical and behavioral heterogeneity. Annals of Internal Medicine, 109, 298-311.

Krauss, M., & Seltzer, M. (1986). Comparison of elderly and adult mentally retarded persons in community and institutional settings. American Journal of Mental Deficiency, 91(3), 237-243.

Lai, F., & Williams, R. (1989). A prospective study of Alzheimer disease in Down syndrome. Archives of Neurology, 46, 849-853.

Libb, J., Myers, G., Graham, E., & Bell, B. (1983). Correlates of intelligence and adaptive behavior in Down's syndrome. Journal of Mental Deficiency Research, 27, 205-210.

Lott, I., & Lai, F. (1982). Dementia in Down's syndrome: Observations from a neurology clinic. Applied Research in Mental Retardation, 3, 233-239.

Mann, D. (1988). The pathological association between Down syndrome and Alzheimer disease. Mechanisms of Ageing and Development, 43, 99-136.

Meyers, C., Nihira, K., & Zetlin, A. (1979). The measurement of adaptive behavior. In N. Ellis (Ed.) Handbook of mental deficiency, psychological theory and research (pp. 431-482). New Jersey: Lawrence Erlbaum Associates.

Miniszek, N. (1983). Development of Alzheimer disease in Down syndrome individuals. American Journal of Mental Deficiency, 87(4), 377-385.

Mitchell, J., Jr. (Ed.). (1985). The ninth mental measurements yearbook. Lincoln: The University of Nebraska Press.

Newroth, A., & Newroth, S. (1980). Coping with Alzheimer's disease: A growing concern. Downsview, Ontario: National Institute on Mental Retardation.

Ninos, M., & Makohon, R. (1985). Functional assessment of the patient. Geriatric Nursing, May/June, 139-142.

Niva, R. (1988). Eye abnormalities and their treatment. In V. Dmitriev, & P. Oelwein (Eds.), Advances in Down syndrome (pp. 45-58). Seattle: Special Child Publications.

Odell, J. (1988). Medical considerations. In C. Tingey (Ed.) Down syndrome: A resource handbook (pp. 33-46). Boston: A College-Hill Publication.

Oliver, C., & Holland, A. (1986). Down's syndrome and Alzheimer's disease: A review. Psychological Medicine, 16, 307-322.

Ortof, E., & Crystal, H. (1989). Rate of progression of Alzheimer's disease. Journal of the American Geriatrics Society, 37(6), 511-514.

Pueschel, S. (1988). Physical characteristics, chromosome analysis, and treatment approaches in Down syndrome. In C. Tingey (Ed.), Down syndrome: A resource handbook (pp. 3-22). Boston: A College-Hill Publication.

Pueschel, S., Tingey, C., Rynders, J., Crocker, A., & Crutcher, D. (1987). New perspectives on Down syndrome, Baltimore: Paul H. Brookes Publishing Co.

Rafalowska, J., Barcikowska, M., Wen, G., & Wisniewski, H. (1988). Laminar distribution of neuritic plaques in normal aging, Alzheimer's disease and Down's syndrome. Acta Neuropathologica, 77, 21-25.

Raynes, N. (1987). Adaptive behavior scales. In J. Hogg, and N. Raynes (Eds.), Assessment in mental handicap: A guide to assessment practices, tests and checklists (pp. 81-106). London: Brookline Books.

Reisberg, B. (1984). Stages of cognitive decline. American Journal of Nursing, 84, 225-228.

Schweber, M. (1985). A possible unitary genetic hypothesis for Alzheimer's disease and Down syndrome. Annals of the New York Academy of Sciences, 450, 223-238.

Schweber, M. (1988). Effects of aging on the development of Alzheimer's disease in adults with Down syndrome. In V. Dmitriev, & P. Oelwein (Eds.), Advances in Down syndrome (pp. 67-74). Seattle: Special Child Publications.

Silverstein, A., Ageno, D., Alleman, A., Derecho, K., Gray, S., & White, J. (1985). Adaptive behavior of institutionalized individuals with Down syndrome. American Journal of Mental Deficiency, 89(5), 555-558.

Silverstein, A., Herbs, D., Nasuta, R., & White, J. (1986). Effects of age on the adaptive behavior of institutionalized individuals with Down syndrome. American Journal of Mental Deficiency, 90(6), 659-662.

Sinex, F. (1986). The mortality of adults with Down syndrome as a function of age. Mechanisms of Ageing and Development, 36, 217-222.

Sinex, F., & Myers, R. (1982). Alzheimer's disease, Down's syndrome, and aging: The genetic approach. In F. Sinex, & C. Merril (Eds.), Alzheimer's disease, Down's syndrome, and aging. Annals of the New York Academy of Sciences, 396, 3-14.

St Clair, D. (1987). Chromosome 21, Down's syndrome and Alzheimer's disease. Journal of Mental Deficiency Research, 31, 213-214.

St Clair,D., & Blackwood,D. (1985). Premature senility in Down's syndrome. Lancet, ii, 34.

Tariot, P., Sunderland, T., Murphy, D., Cohen, R., Weingartner, H., & Makohon, R. (1985). How memory fails: A theoretical model. Geriatric Nursing, May/June, 144-147.

Thase, M. (1982). Longevity and mortality in Down's syndrome. Journal of Mental Deficiency Research, 26, 177-192.

Thase, M. (1988). The relationship between Down syndrome and Alzheimer's disease. In L. Nadel (Ed.), The psychobiology of Down syndrome, (pp. 345-368). London: The MIT Press.

Thase, M., Smeltzer, L., & Maloon, J. (1982). Clinical evaluation of dementia in Down's syndrome: A preliminary report. Journal of Mental Deficiency Research, 26, 239-244.

Tukey, J. (1977). Exploratory data analysis. Don Mills: Addison-Wesley Publishing Company.

Winogrond, I., Fisk, A. (1983). Alzheimer's disease: Assessment of functional status. Journal of the American Geriatrics Society, 31(12), 780-785.

Wishart, J. (1988). Early learning in infants and young children with Down syndrome. In L. Nadel (Ed.), The psychobiology of Down syndrome, (pp. 7-50). London: The MIT Press.

Wisniewski, K., Dalton, A., Crapper-McLachlan, D., Wen, G., & Wisniewski, H. (1985). Alzheimer's disease in Down's syndrome: Clinicopathologic studies. Neurology, 35, 957-961.

Wisniewski, H., & Rabe, A. (1986). Discrepancy between Alzheimer-type neuropathology and dementia in persons with Down's syndrome. Annals of the New York Academy of Sciences, 477, 247-259.

Wisniewski, K., Wisniewski, H., & Wen, G. (1985). Occurrence of neuropathological changes and dementia of Alzheimer's disease in Down's syndrome. Annals of Neurology, 17, 278-282.

Wright, A., & Whalley, L. (1984). Genetics, aging, and dementia. British Journal of Psychiatry, 145, 20-37.

Zigman, W., Schupf, N., Lubin, R., & Silverman, W. (1987). Premature regression of adults with Down syndrome. American Journal of Mental Deficiency, 92(2), 161-168.

Appendix A

Correspondence

March 1, 1990

(name of chairperson), Chairperson
Michener Centre Research Sub-Committee
Michener Centre
Box 5002
Red Deer, Alberta, T4N 5Y5

Dear Mrs. (name of chairperson),

I am completing a Master's degree in Educational Psychology at the University of Alberta. At the present time I am working on my thesis under the supervision of Dr. Dick Sobsey (492-3755). I have attached for your consideration:

1. an Application for Conducting Research at Michener Centre,
2. an Ethical Guidelines Checklist,
3. my Research Proposal for the project.

Please accept this as a formal request for approval to conduct research at Michener Centre, in order to collect the data necessary to complete my Master's thesis.

The topic I have chosen to study is: Effects of Age on the Adaptive Behavior of Institutionalized Down Syndrome Adults. My proposal presents the background and rationale of my topic and the research question to be investigated. In addition, data collection procedures, and a consideration of how the anonymity of subjects and confidentiality of information will be protected is described. The proposal was approved by Dr. Sobsey at the University of Alberta on February 27, 1990. It has been submitted to the University Ethics Committee for their approval.

Thank you for your consideration of my request.

Sincerely,

Dianne Rasmussen

Alberta

SOCIAL SERVICES

MEM[illegible]U

FROM Elaine Saunders, Chairperson
Michener Centre
Research Sub-Committee

OUR FILE REFERENCE

YOUR FILE REFERENCE

TO Dianne Rasmussen
Staff Development
Michener Centre

DATE March 30, 1990

TELEPHONE 5619

SUBJECT RESEARCH PROPOSAL APPLICATION

Your proposal entitled "Effects of Age on the Adaptive Behavior of Institutionalized Down Syndrome Adults" has been approved for implementation.

May we request on-going consultation with Mr. Ho, Psychological Services to maintain effective communication regarding your project. The Sub-Committee will be requesting a progress report on results achieved October 31, 1990.

On behalf of the Committee members, Dr. S. Koop and Mr. D. Verstraete, may I wish you success in your thesis project.

Elaine Saunders

Elaine Saunders, Chairperson
Michener Centre
Research Sub-Committee

ES/cls

FROM: Dianne Rasmussen
Staff Development
Michener Centre

March 31, 1990

TO: Elaine Saunders, Chairperson
Michener Centre
Research Sub-Committee

SUBJECT: Research Proposal Approval

I would like to thank the members of the Michener Centre Research Sub-Committee for their review and approval of my research project entitled "Effects of Age on the Adaptive Behavior of Institutionalized Down Syndrome Adults".

As the proposal has also been approved by the University of Alberta Ethics Committee, I plan to begin the project immediately. I would be pleased to consult with Mr. Ho on an on-going basis, and provide a progress report to the Research Sub-Committee by October 31, 1990.

Once again, thank you for your consideration.

Dianne Rasmussen
Staff Development
Michener Centre

Alberta **MEMORANDUM**

FAMILY AND SOCIAL SERVICES
~~AGRICULTURE~~

MICHENER CENTRE RED DEER

FROM JAMES O. YOUNG
Director - Records Management

OUR FILE REFERENCE

YOUR FILE REFERENCE

TO DIANNE RASMUSSEN
STAFF DEVELOPMENT CO-ORDINATOR

DATE 90.04.05

TELEPHONE 5769

SUBJECT ACCESS TO CLIENT FILES

You have been cleared to have access to client files for your research.

As you are aware these records are very confidential and you must not use any client names in your paper.

If you choose to use these after hours you are to check in at the switchboard to get the key and return it to switchboard as you leave.

Good luck with your project!

Jim

JAMES O. YOUNG
DIRECTOR - RECORDS MANAGEMENT

JOY/hr

cc.
Jim McCormick - Director - Social Services
Switchboard
Helga Ross - Supervisor - Central Client Records

Appendix B

Data Form

Data Form

Admission Date: _____ I.D. #_____

Gender: M F Group: DS non-DS

<40 40+

M/M S/P

THE PYRAMID SCALES:

Date of Administration:	(1)_____	(2)_____
Birthdate:	_____	_____
C.A. at Administration:	_____	_____
Functioning Level:	_____	_____
Residential Unit:	_____	_____

Subdomain	Percentage	Percentage
I. Sensory Zone		
1. Tactile Responsiveness	_____	_____
2. Auditory Responsiveness	_____	_____
3. Visual Responsiveness	_____	_____
II. Primary Zone		
4. Gross Motor	_____	_____
5. Eating	_____	_____
6. Fine Motor	_____	_____
7. Toileting	_____	_____
8. Dressing	_____	_____
9. Social Interaction	_____	_____
10. Washing/Grooming	_____	_____
11. Receptive Language	_____	_____
12. Expressive Language	_____	_____
III. Secondary Zone		
13. Recreation/Leisure	_____	_____
14. Writing	_____	_____
15. Domestic Behavior	_____	_____
16. Reading	_____	_____
17. Vocational	_____	_____
18. Time	_____	_____
19. Numbers	_____	_____
20. Money	_____	_____
Total Score	_____	_____
Mean	_____	_____

Appendix C

Supplementary Tables

Table A Median (Md) and Interquartile Range (IQR) Percentage Competency per Subdomain as a Function of Etiology, Age, and Functioning Level

	Mild/Moderate				Severe/Profound			
	<40[a]		40+[b]		<40[c]		40+[d]	
Subdomain	Md	IQR	Md	IQR	Md	IQR	Md	IQR
Sensory-Motor								
TR								
DS	100	0	100	9.0	88	28.0	81	35.0
non-DS	100	0	100	9.0	88[e]	25.0	88	22.0
AR								
DS	100	6.0	94	25.0	75	33.5	66	36.5
non-DS	97	12.0	94[f]	12.0	75[e]	32.0	75[g]	27.2
VR								
DS	100	12.0	91	25.0	63	32.5	63	37.5
non-DS	89	22.0	94	25.0	63	22.0	66	31.0
GMOT								
DS	100	3.0	97	4.5	91	13.0	88[g]	19.0
non-DS	100	7.5	97	35.5	94	19.0	88	13.0
FMOT								
DS	94	6.0	91	8.0	59[e]	28.0	63	26.5
non-DS	91	13.0	94	13.0	59	39.5	59	27.5
Self-Help								
EAT								
DS	97	4.5	97[f]	2.3	72	32.0	73.5[g]	32.5
non-DS	97	7.5	94	15.5	72	32.0	69	26.0
TOIL								
DS	100	0	100	1.5	84	42.5	89	32.5
non-DS	100	3.0	100	7.5	78	53.0	75	26.5
DRES								
DS	100	3.0	100	3.0	69	29.5	69	45.5
non-DS	94	12.5	97	10.0	63	50.0	63	32.5
WASH								
DS	100	6.0	100	4.5	50	50.0	59	56.0
non-DS	91	20.5	97	19.0	47	51.5	44	41.0

Table A Continued

	Mild/Moderate				Severe/Profound			
	<40[a]		40+[b]		<40[c]		40+[d]	
Subdomain	Md	IQR	Md	IQR	Md	IQR	Md	IQR
Communication								
RLAN								
DS	84	16.0	81	16.0	44	34.0	44	50.0
non-DS	88[h]	12.3	89.5[f]	18.3	44	44.0	38	41.0
ELAN								
DS	94	17.3	97	26.5	19	25.0	31	45.0
non-DS	94	16.0	94	23.0	22	60.0	38	68.0
Socialization								
SOCI								
DS	84	16.0	75	28.0	41	28.0	44	43.5
non-DS	78	22.0	81	23.5	44	25.0	38	28.0
REC								
DS	72	20.5	66	22.0	25	32.0	22	31.5
non-DS	57	29.5	53	23.0	28	27.0	22	15.0
DOME								
DS	81[h]	15.5	75	33.5	13	28.0	19	30.0
non-DS	56	44.0	50	63.5	13	25.0	13	17.5
VOC								
DS	61[i]	16.0	57.5[f]	12.2	0	22.0	0	14.0
non-DS	50[j]	20.0	51.5[f]	26.5	0	22.0	0	0
Cognitive								
WRIT								
DS	54.5[h]	15.0	44	16.5	19	20.0	22	23.5
non-DS	53	21.0	53	22.0	19	19.0	22	25.0
READ								
DS	25	26.5	22	3.0	3	16.0	6	13.0
non-DS	25	22.5	25	37.5	6	13.0	3	11.5
TIME								
DS	50	41.0	47	31.0	6	11.0	3	16.0
non-DS	50	42.0	44	59.5	0	13.0	3	10.5

Table A Continued

Subdomain	Mild/Moderate				Severe/Profound			
	<40[a]		40+[b]		<40[c]		40+[d]	
	Md	IQR	Md	IQR	Md	IQR	Md	IQR
NUM								
DS	38	28.0	25	21.5	0	3.0	0	4.5
non-DS	31	28.0	25	37.0	0	3.0	0	1.5
MON								
DS	31	31.0	19	18.0	0	9.0	0	3.0
non-DS	22	18.0	16	39.0	0	7.5	0	4.5
Mean Score								
DS	76	12.5	72	9.5	39	24.5	43	24.5
non-DS	73	13.5	70	22.0	44	26.5	37	23.0

$a_n = 37$ $b_n = 13$ $c_n = 61$ $d_n = 29$
$e_n = 60$ $f_n = 12$ $g_n = 28$ $h_n = 36$ $i_n = 34$ $j_n = 33$

Table B Mean (M) and Standard deviation (SD) Percentage Competency per Domain as a Function of Etiology, Age, and Functioning Level

	Mild/Moderate				Severe/Profound			
	<40[a]		40+[b]		<40[c]		40+[d]	
Domain	M	SD	M	SD	M	SD	M	SD
Sensory-Motor								
DS	94.6	6.3	92.5	5.1	71.6	14.2	68.9	18.1
non-DS	92.5	6.7	91.4	6.8	70.9	17.6	72.8	13.7
Self-Help								
DS	97.8	1.8	98.2	1.9	64.2	21.4	67.3	24.4
non-DS	92.4	6.9	91.6	9.8	59.8	25.3	61.6	19.2
Communication								
DS	84.7	14.5	82.4	16.7	35.2	20.7	40.4	27.1
non-DS	85.9	15.0	81.7	25.2	40.5	26.5	43.5	28.3
Socialization								
DS	73.6	9.7	68.6	8.8	24.8	16.3	25.2	17.4
non-DS	61.5	14.5	58.8	18.0	24.3	15.9	20.0	10.6
Cognitive								
DS	42.2	16.7	31.8	10.9	8.1	7.2	9.2	8.5
non-DS	39.8	19.7	38.9	22.4	8.7	8.8	7.8	8.5

[a] n = 37 [b] n = 13 [c] n = 61 [d] n = 29 per group

Table C Mean (M) and Standard deviation (SD) Percentage Competency per Subdomain as a Function of Etiology, Age, and Functioning Level

	Mild/Moderate				Severe/Profound			
	<40[a]		40+[b]		<40[c]		40+[d]	
Subdomain	M	SD	M	SD	M	SD	M	SD
Sensory-Motor								
TR								
DS	97.6	6.2	96.4	5.8	83.9	14.7	79.8	19.2
non-DS	97.8	5.5	96.7	5.2	85.3[e]	15.1	85.0	13.8
AR								
DS	94.3	12.0	89.7	11.3	71.8	20.4	65.7	25.3
non-DS	91.7	11.4	92.4[f]	7.5	70.7[e]	20.8	74.8[g]	19.0
VR								
DS	90.0	20.1	87.8	12.2	55.7	26.0	53.8	26.3
non-DS	89.4	11.5	90.0	12.0	58.7	24.6	57.5	30.3
GMOT								
DS	98.1	2.9	97.4	2.4	88.3	10.0	86.3[g]	13.4
non-DS	95.0	8.7	87.3	17.7	85.3	18.1	86.0	11.6
FMOT								
DS	92.9	5.2	91.0	5.3	58.6[e]	18.5	58.4	21.3
non-DS	88.6	9.9	90.1	9.0	55.9	24.7	60.8	17.9
Self-Help								
EAT								
DS	96.2	3.9	97.0[f]	2.6	71.6	17.9	69.2[g]	23.6
non-DS	93.4	7.1	91.0	9.3	69.1	21.3	71.1	14.1
TOIL								
DS	99.6	0.9	98.8	2.6	75.1	25.0	78.3	26.1
non-DS	98.1	3.5	95.3	9.8	69.1	29.7	70.4	25.7
DRES								
DS	97.9	2.6	98.6	2.3	62.2	24.2	63.3	27.4
non-DS	90.4	10.3	91.1	10.7	55.5	28.9	58.0	23.1
WASH								
DS	97.4	3.6	97.9	3.6	47.7	26.0	57.5	30.8
non-DS	87.6	11.3	89.0	16.3	45.3	27.8	46.5	23.2

Table C Continued

Subdomain	Mild/Moderate <40[a] M	Mild/Moderate <40[a] SD	Mild/Moderate 40+[b] M	Mild/Moderate 40+[b] SD	Severe/Profound <40[c] M	Severe/Profound <40[c] SD	Severe/Profound 40+[d] M	Severe/Profound 40+[d] SD
Communication								
RLAN								
DS	82.6	11.5	80.1	12.6	41.5	21.7	41.8	29.5
non-DS	85.7[h]	10.3	85.6[f]	14.4	43.4	24.5	41.5	25.4
ELAN								
DS	86.7	19.0	84.6	22.9	28.9	22.9	38.8	28.0
non-DS	87.8	17.5	83.3	28.0	37.5	31.1	45.4	34.5
Socialization								
SOCI								
DS	84.2	11.3	79.6	15.7	42.2	19.1	46.9	24.7
non-DS	79.2	14.0	78.4	14.9	42.6	19.5	41.1	17.5
REC								
DS	70.0	13.4	63.1	14.1	26.8	18.4	25.7	18.0
non-DS	58.5	16.3	58.9	17.2	28.5	18.2	24.6	13.1
DOME								
DS	79.6[h]	14.0	72.2	16.3	17.9	21.1	17.0	15.4
non-DS	58.5	22.9	47.3	34.3	16.1	17.7	10.8	11.5
VOC								
DS	60.4[i]	11.3	58.4[f]	7.1	12.0	18.0	11.0	20.3
non-DS	48.9[j]	14.6	49.4[f]	17.3	10.0	17.2	3.4	9.5
Cognitive								
WRIT								
DS	55.5[h]	12.1	46.0	8.9	17.9	13.9	21.1	15.8
non-DS	51.8	18.4	55.6	17.1	19.5	15.1	18.6	15.7
READ								
DS	33.0	22.0	22.0	9.0	7.1	8.5	7.7	7.2
non-DS	32.7	23.5	30.7	24.9	8.4	9.0	6.2	7.2
TIME								
DS	53.1	25.8	43.9	19.7	8.0	9.7	9.4	12.5
non-DS	53.1	25.0	50.3	32.4	7.2	10.3	7.4	11.7
NUM								
DS	37.2	17.1	24.6	14.1	2.8	5.7	4.8	9.7
non-DS	32.6	20.3	30.2	21.9	4.4	9.0	3.2	7.6

Table C Continued

	Mild/Moderate				Severe/Profound			
	<40[a]		40+[b]		<40[c]		40+[d]	
Subdomain	M	SD	M	SD	M	SD	M	SD
MON								
DS	33.0	19.7	22.4	10.2	4.4	6.5	2.6	5.3
non-DS	28.4	18.8	28.0	26.2	3.8	6.7	3.4	6.7
Mean Score								
DS	77.1	7.7	72.1	5.3	41.2	13.6	41.9	16.0
non-DS	73.3	10.2	70.7	12.9	40.8	15.9	40.7	12.0

[a] n = 37 [b] n = 13 [c] n = 61 [d] n = 29
[e] n = 60 [f] n = 12 [g] n = 28 [h] n = 36 [i] n = 34 [j] n = 33