PROTECTIVE FACTORS FOR THE DEVELOPMENT OF ALZHEIMER'S DEMENTIA IN ADULTS WITH DOWN SYNDROME

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ABSTRACT

PROTECTIVE FACTORS FOR THE DEVELOPMENT OF ALZHEIMER'S DEMENTIA IN ADULTS WITH DOWN SYNDROME

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This study compared adults with Down syndrome who developed symptoms of Alzheimer's dementia with those who remained symptom-free in order to uncover some of the factors associated with decline. The focus was on factors which may be protective against the development of dementia in individuals with Down syndrome such as high level of cognitive functioning, duration of education, type of employment, and involvement in various recreational activities and hobbies. The effects of anti-inflammatory drugs and smoking were also explored. Results indicate that greater age and higher level of cognitive function were the only variables associated with decline. Discussion focused on the factors which may influence an individual's level of cognitive functioning.

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Introduction

Improving our understanding of Alzheimer's disease (AD) is now becoming a national priority given the anticipated threefold increase in the number of cases expected in the coming years (National Institute of Aging, 1980). Research on this issue is now underway in many centres around the world, and one group of individuals who will likely be the subject of some of this research is adults with Down syndrome. This is because it is now well established that a large percentage of adults with Down syndrome who live to adulthood develop the neuropathology of AD (Holland, 1995; Mann, 1988; Zigman, Silverman, & Wisniewski, 1996). Jervis (1948) was the first to discover the relationship between the two disorders when he found AD pathology in his autopsies of three adults with Down syndrome. Since that time, a large amount of research has confirmed the unique nature of the relationship between Down syndrome and AD (e.g., Evenhuis, 1990; Karlinsky, Holland & Berg, 1993; Mann, 1988). It is currently believed that the causes of AD may be related to the causes of Down syndrome, and that we may be able to discover important information about both disorders through their combined study (Holland, 1994).

One of the most intriguing aspects of AD in Down syndrome is that although the vast majority of individuals over 40 years of age develop the AD neuropathology, only about 50% of them display the clinical syndrome of dementia (Holland, 1994; Zigman et al., 1993). This suggests that genetic factors associated with chromosome 21 and Down syndrome may not be sufficient to account for the onset of AD in Down syndrome individuals and that research into environmental, social and constitutional factors may be in order for a more complete understanding of the onset of AD for this group.

The aim of the present study was to explore whether some of the protective factors thought to exist for AD in the general population would apply to the Down syndrome population as well. Research of this kind may help explain why only 50% of individuals with Down syndrome develop the dementing syndrome while virtually all are found to have the pathology. Factors investigated in this study included years of education, residential placement, level of recreational activity (e.g. hobbies, sports), type of employment, and degree of intellectual functioning. As well, the use of nonsteroidal anti-inflammatory drugs and cigarette smoking was investigated on a preliminary basis. By comparing the past and present environments, activities, and attributes of those who exhibit decline with those who remain healthy, it may be possible to uncover factors important in dementia's onset.

Down Syndrome and Alzheimers Disease

Down syndrome is a genetic disorder resulting from abnormalities in chromosome 21 and occurring in approximately 1 in 600 live births (Holland, 1994). This makes it one of the most common known causes of mental retardation. As well as mental retardation, individuals with Down syndrome suffer from a number of physical and health problems including an increased incidence of congenital heart disease, hypothyroidism, seizures, and degenerative vascular disease (Zigman et al., 1996). As a result of these medical complications, the lifespan of individuals with Down Syndrome has, in the past, been shorter than average (Karlinsky, Holland & Berg, 1993; Strauss & Eyman, 1996; Zigman, et al., 1996). However, recent improvements in medical technology have allowed individuals with Down syndrome to live longer. Currently, the average lifespan for a 1-year-old individual with Down syndrome and mild retardation is 55 years (Strauss & Eyman, 1996).

Alzheimer's Disease is a neuropathological disorder which results in a

number of changes in the brain, the most cytoanatomically relevant being the formation of senile plaques and neurofibrillary tangles (Gottfries, Blennow, Gottfries, Karlsson & Wallin, 1995). Though its presenting features are quite variable, the clinical expression of AD generally has an insidious onset and a slow but progressive course. Symptoms commonly experienced in the initial stages of the disease by those without Down syndrome include memory loss for recent events, word finding difficulties, indecisiveness, difficulty acquiring new information, and some cognitive disorientation (Alzheimer's Society, 1991; DSM-IV, 1994). By the final stages, individuals lose "virtually all intellectual capacities" and experience severe motor deterioration (Karlinsky, et al., 1993, p. 5). Guidelines contained in the DSM-IV (1994) for the diagnosis of AD include the presence of memory impairment, disturbances in abstract thinking and judgment, disturbances in language functions, impaired motor function, and failure to recognize or identify familiar objects (agnosia).

The course of AD in the Down syndrome population is not as clear. The limited expressive capabilities of adults with Down syndrome often make the early, less conspicuous signs of dementia, such as word finding deficits, difficult to detect (Wisniewski & Silverman, 1996). As well, adults with Down syndrome often live in supported environments (e.g. group homes) where many of the demands of daily living are handled by support workers (Brown, Raphael, & Renwick, 1997). As a result, adults with Down syndrome often engage in fewer activities in which decline could be seen, making detection of the first signs of AD more difficult. This can be especially true of those with lower levels of functioning (Aylward, Burt, Thorpe, Lai & Dalton, 1997). Because initial detection may be problematic, it is conceivable that what we describe as the first symptoms of AD in individuals with Down syndrome are actually symptoms of a more advanced stage. At

present, documented first signs of AD include extended periods of apathy, loss of skills, memory problems, disorientation, loss of speech, and various abnormal neurological signs (Byrd-Burt, Loveland, Kay & Lewis, 1992; Lai & Williams, 1989; Zigman et al., 1993). As well, late-onset seizures often develop. Lai & Williams (1989) reported that 84% of the adults with Down syndrome referred to them, due to behaviour or cognitive decline, developed seizures within an eight year period.

Evidence Linking Down syndrome and Alzheimer's disease

Virtually all adults with Down syndrome over the age of 40 develop neuropathology consistent with AD (Holland, 1994; Lai & Williams, 1989; Mann, 1988; Zigman et al. , 1993). In his comprehensive meta-analysis of 39 studies involving autopsies of individuals with Down syndrome, Mann (1993) found that of 211 patients with Down syndrome over 40 years of age, only three did not exhibit AD pathology upon autopsy. Further comparison of the morphology and microchemistry of the plaques and tangles in Down syndrome and AD revealed only slight differences between the two. Mann suggested these variations were likely due to differences in disease duration. He concluded that "...in pathological terms, patients with Down's syndrome at middle age do indeed have Alzheimer's disease" (Mann, 1988, p. 125).

Epidemiological research provides another line of evidence for the association of Down syndrome and AD. The Canadian Study of Health and Aging (1994) found that a positive family history of mental retardation was more frequent among those diagnosed with AD, and although they attempted to examine Down syndrome specifically, the number of families with a Down syndrome member was too small to examine further. More convincing epidemiological results come from the meta-analysis conducted by the EURODEM (Vanduijn, Stijnen, Hofman et. al., 1991) group. EURODEM is

an international group of researchers who reanalyzed the results of seven different studies from Australia, Italy, the Netherlands and the U.S.A. and found a strong and significant relationship between the development of AD and the incidence of Down syndrome in first degree relatives. This finding suggests a familial link between AD and Down syndrome. Given the large number of subjects included in this analysis (over 800 in all) and the cross-cultural nature of the EURODEM study, these findings can be viewed with a fair degree of confidence.

One final but compelling line of evidence for the association between AD and Downs syndrome comes from genetic studies. Down syndrome is, in over 95% of cases, the result of inheriting an extra chromosome 21, a condition commonly referred to as trisomy 21 (Holland, 1994). Genetic research has also found chromosome 21 to be important in the development of AD. Researchers at the University of Toronto (St. George-Hyslop et al. , 1987) reported linkage to a chromosome 21 marker in some families developing early onset AD. These findings suggest that chromosome 21 may be a good "candidate gene" for finding the locus of some types of AD as well as for Down syndrome (Holland, 1995).

The Clinical Diagnosis of AD

Diagnosing AD in the Down syndrome population has been problematic for a number of reasons. Because adults with Down syndrome vary greatly in their intellectual abilities it is difficult, if not impossible, to use established test norms to detect decline. Each individual with Down syndrome has a different pattern of cognitive strengths and weaknesses and this makes test norms meaningless. In order to document decline in functioning for adults with Down syndrome, it is necessary to have individual "baseline" data on each individual's level of cognitive

functioning. For this reason, it is desirable to administer a battery of psychological tests to all adults with Downs syndrome in early adulthood, before decline starts. If this is done, records describing optimal level of function will exist and can be compared to later levels if concern regarding decline arises (Aylward, Burt, Thorpe, Lai & Dalton, 1997).

Another problem with diagnosing AD in those with Down syndrome is the great range of ability that is found in these individuals. Those with a mild level of delay are able to perform on psychometric tests, however those in the more severe range of delay are frequently unable to do so. In practice, this results in formal testing being performed to diagnose the mildly delayed, while caregiver reports are used for assessing the severely delayed. For research purposes, this may mean that there is no uniformity of assessment in terms of instrumentation since no single instrument will be appropriate for all individuals with Down syndrome. As well, information from caregivers can be particularly difficult to evaluate, in terms of reliability and validity.

Recently, the American Association on Mental Retardation (AAMR) and the International Association for the Scientific Study of Intellectual Disability (IASSID) published guidelines proposing a standardized methodology for the diagnosis of AD in Down syndrome (Aylward, Burt, Thorpe, Lai & Dalton, 1997). Their suggestions included criteria for the diagnosis of AD in Down syndrome, and a number of tests and interview protocols that might be used for this purpose. The group stated a preference for the use of the International Classification of Diseases (ICD-10) criteria for diagnosis since it places a greater emphasis on the behavioural manifestations of dementia. As well, the group believed that the ICD-10 method of diagnosing dementia alone first, and then moving to a specific

diagnosis of AD, was superior to the DSM-IV method. Criteria for dementia in ICD-10 include: (a) a decline in memory, such as an inability to remember social arrangements, (b) a decline in other cognitive functions, such as agnosia, apraxia or aphasia, (c) a loss of emotional control, motivation or social behaviour, such as increased irritability, and (d) a duration of symptoms lasting at least 6 months. The guidelines go on to describe other possible conditions that may lead to dementia that must be excluded before a diagnosis of AD can be given. These include hypothyroidism, depression, and medication-induced dementia.

Onset of the Clinical Syndrome of Dementia

As indicated earlier, although the vast majority of adults with Down syndrome develop AD neuropathology, fewer develop the clinical signs of dementia. Estimates of the percentage of those who develop clinical dementia vary from a low of 0%, in a community sample (Devenny et al., 1996), to a high of 88% in an institutionalized sample (Evenhuis, 1990). The majority of studies, however, report that fewer than 50% of adults with Down syndrome, over 40 years of age, develop significant signs of dementia (Zigman et al., 1993).

The variation in reported prevalence rates described above suggests that factors other than chromosome 21 abnormality, or other genetic causes, may be involved in determining either the age of onset of dementia in Down syndrome, or whether a dementing syndrome will develop at all. Epidemiological studies in the non-Down syndrome population have revealed a number of risk and protective factors that are believed to be associated with the onset of AD, and which may influence disease onset. These factors may prove to be of relevance in predicting the onset of dementia in the Down syndrome population as well.

Factors Which May Effect the Onset of Alzheimer's disease

An extensive literature exists regarding risk and protective factors for AD in the general population. Among the most often reported protective factors is level of education (Canadian Study of Health and Aging, 1994; Katzman, 1993). A number of studies have found a negative correlation between education and dementia, where those with higher levels of education are less likely to develop dementia (Friedland, 1993; Katzman, 1993; Canadian Study of Health and Aging, 1994). Katzman (1993) has suggested that higher levels of education reflect what he describes as high "neuronal reserve". High neuronal reserve could be the outcome of more years of education and reflect environmental enrichment, or it could be the reason why higher levels of education are attained in the first place, due to a greater genetic endowment. In either case, neuronal reserve is presumed to reflect synaptic density and increased brain weight. When AD degeneration begins, a person with high levels of neuronal reserve should present more resistance to deterioration, since he/she will have a greater neuronal capacity upon which to draw. This could result in a delay of dementia onset and, given the advanced age at which AD begins, a patient may die of other causes prior to clinical symptom onset. In other words, having greater neuronal reserve would not prevent the disease, but it may defer expression long enough to appear to prevent it.

Swaab (1991) described how increased environmental stimulation in rats resulted in a greater cortical weight, size and thickness, as well as increased dendritic branching. Rats experiencing enhanced environmental stimulation also showed better performance on complex maze tasks than their less stimulated counterparts. These results with lower animals, which are presumed to generalize to humans, suggest that environmental

enrichment, such as education or cognitively challenging occupations or activities, should result in greater brain size, weight and thickness, and therefore a larger "neuronal reserve" with which to combat AD. They also suggest a connection between synaptic density and greater ability or functional intelligence.

Autopsy studies provide an interesting source of evidence for the role of neuronal reserve in AD. Katzman (1993) identified a group of non-Down syndrome subjects with a sufficient quantity of senile plaques to meet criteria for AD, but who did not experience dementia. These subjects differed from other demented subjects with similar quantities of plaques in that they had a "greater number of large neurons in the parietal cortex", and a greater overall brain weight (Katzman, 1993, p. 18). Katzman suggests that these subjects' greater neuronal reserve (increased brain weight and number of neurons) protected them from developing the clinical signs of dementia. It is interesting to note that the presence of plaques and tangles without a dementing syndrome is reminiscent of the situation in adults of Down syndrome.

A second line of evidence for the effect of neuronal reserve comes from tests of cerebral blood flow in patients with AD. Stern et al. (1992) demonstrated that when patients were matched for symptom severity, those with greater education, and presumably greater neuronal reserve, showed diminished blood flow to the tempoparietal cortex. Since reduced blood flow suggests greater pathology, the more educated group appeared to manifest a similar degree of symptom severity, but with greater cortical damage. This suggests that neuronal reserve, measured through higher education, may be protecting subjects from the effects of AD degeneration.

A second factor which may protect against the development of AD is

exposure to cigarette smoke. In his meta-analysis of 19 studies of non-Down syndrome individuals, Lee (1994) found that the risk for AD is lower for those with a history of cigarette use. There is also evidence that nicotine improves performance on cognitive tasks in normal elderly patients (Wesnes & Warburnton, 1984), and that nicotinic receptor binding sites are reduced in AD (Lee, 1994). Together, this evidence suggests that those who smoke or who are exposed to cigarette smoke, may be protected against AD. This, interestingly, suggests that brain stimulation of any kind, including the type that results in compromising the health of typical individuals, may act as a protective factor for AD much as education has been shown to do.

The use of anti-inflammatory drugs has also been proposed as a protective factor for AD. The Canadian Study of Health and Aging (1994) found that subjects who had a history of anti-inflammatory use, or of arthritis, were found to develop AD less often. As anti-inflammatories are often prescribed for arthritis, their common relationship to AD provides convergent evidence for this factor. It is currently believed that anti-inflammatories reduce the expansion of harmful substances in the brain and this defers the destruction of neurons (Goldberg, 1996).

Objectives of this Study

In line with Katzman's (1993) theory of neuronal reserve, environmental enrichment might be presumed to result in cognitive enrichment. This, in turn, should result in a deferral of dementia symptoms. For individuals with Down syndrome factors such as number of years of education, number of recreational activities, and employment activity may be good indicators of overall environmental stimulation. As well, assuming that community living is more cognitively challenging than institutional life, fewer years spent in an institution should also indicate greater environmental enrichment. Using these indicators, this study compared individuals with Down syndrome who were found to have

AD to those who remained symptom free in order to establish if the two groups differed. Specifically, this study investigated whether individuals with (a) more years of education, (b) more challenging employment, (c) more recreational activities, and (d) fewer years in an institution, would experience less decline.

Hereditary or congenital factors could also have an impact on neuronal reserve. Children with Down syndrome born with a mild to moderate level of retardation could be seen as having higher levels of innate neuronal reserve. According to Katzman's (1993) theory, this group might be expected to experience less decline than those with severe to profound retardation. To clarify whether there is support for the hypothesis that level of cognitive functioning is systematically related to onset of decline in individuals with Down syndrome, this study examined the level of functioning of those developing dementia and those remaining healthy. It was hypothesized that those with a lower level of functioning would experience more decline than those with a higher level of functioning. The last aim of this study was to conduct a preliminary investigation into the links between anti-inflammatory drugs, cigarette smoking and decline in Down syndrome to possibly clarify whether these factors influence disease onset.

Figure 1 shows the hypothetical relationship between variables. It was predicted that each variable would be independently related to the onset of decline, and that each would contribute some unique variation to symptoms of AD. Although it would also have been desirable to test the hypothesis that lower level of functioning and a less stimulating environment result in the onset of AD symptoms earlier in life, it was not possible to do so in the context of this study. The limited time period of this project made it impossible to follow participants through on a longitudinal basis.

Method

Participants

Participants were adult clients with Down syndrome involved in receiving baseline psychological assessments from the Down syndrome Service at Surrey Place Centre in Toronto. Surrey Place Centre is a facility for the assessment and treatment of people with developmental disabilities in the Metropolitan Toronto area. During the course of the study, attempts were made to contact all clients over the age of 35 previously seen at the Down syndrome Service for follow-up assessment and inclusion in the study. As well, all new clients referred to psychological services in the six month time span of the study due to concerns regarding a possible cognitive decline were invited to participate.

Of the 38 clients over 35 years of age who previously received services from the Down syndrome Clinic, two could not be located due to change of residence, and another seven refused services at Surrey Place Centre, leaving 29 eligible participants. Of this 29, two individuals had died and one had been placed in a nursing home. Both the deceased clients and the "nursing home" client had been previously diagnosed with AD by at least two separate clinicians and their data were included in the analysis using information from their records. All remaining clients were seen by the facility's psychological staff and agreed to participate in the study.

Over the six month period of the study, 13 clients were referred to psychological services due to suspected decline. Of these, one was excluded from the study due to a diagnosis of normal pressure hydrocephalus, another one was excluded due to the recent removal of a brain tumor, and four were excluded due to insufficient background information for analysis (e. g. no clinical records available and no caregiver knowledgeable enough to inform).

In total, seven new participants were included bringing the total number of participants to 36.

Procedure

When clients arrived for their assessments, the study was explained to them and they were offered the opportunity to participate (see Appendix 1 for Client and Caregiver Information Sheets and Appendix 2 for Client and Caregiver consent forms). Participation involved allowing the experimenter to view client records and have access to current psychological test scores for the purposes of assigning a diagnosis of decline or no decline. Clients were also asked to allow their caregivers to complete questionnaires concerning the recreational activities and general health of the client [see Appendix 3 for the Health and Activities Questionnaire and the revised Residential Lifestyles Inventory (Kennedy, Horner, Newton, & Kanda (1990)]. All consenting clients received the standard neuropsychological battery presently used at Surrey Place Centre to assess level of decline in Down syndrome. This battery was administered by the centre's neuropsychologist or an experienced psychometrist, Dr. E. Jozsvai or Ms. B. Dunleavy, respectively, using standard psychometric procedures for this population.

Screening Methods

In order to screen for thyroid dysfunction, visual or hearing deficits, sleep apnea, medication induced decline, stroke and depression, all of which are possible alternate causes of cognitive or functional decline, the Differential Diagnosis Screening Questionnaire contained in the Dementia Scale for Down Syndrome (DSDS; Gedye, 1995) was administered to all caregivers. Clients suspected of having hearing deficits were asked to accept appointments with Surrey Place's audiologist. Where results suggested the possibility of thyroid dysfunction, clients were asked to undergo medical tests

by their physicians, as necessary. All other causes of dementia suggested by this questionnaire were investigated and differential diagnoses made.

Instruments

Independent measures. Primary caregivers for each client were asked to complete the Residential Lifestyles Inventory (RLI; Kennedy, Horner, Newton & Kanda, 1990) and the Health and Activities questionnaire (see Appendix 3). The RLI was used to provide details as to the client's social and leisure activities and allow comparison among clients. Originally, the RLI was created to study client preference for various activities rather than simply measure level of activity. In revising it for use in this study, questions regarding level of preference were dropped and a simpler scale applied. The Health and Activities questionnaire provided detailed information concerning the client's (a) educational history, (b) health, including use of prescription and/or other over-the-counter drugs (e.g. nonsteroidal anti-inflammatories), (c) cigarette smoking, (d) occupational history, and (e) residential history. Additional information in these areas was also obtained through review of client records at Surrey Place.

Premorbid level of cognitive functioning was determined by dividing participants into 5 categories, based on DSM-IV (1994) criteria for mild, moderate, and severe delay. The 5 categories were: (a) mild, with an IQ of 55 or greater (b) mild/moderate, with an IQ of 49-54 (c) moderate, with an IQ of 43-48 (d) moderate/severe, with an IQ of 37-42 and (e) severe/profound, with an IQ of less than 36. Information regarding participants' level of cognitive functioning was gathered from clinical records. In 16 cases, it was possible to get IQ scores from records of previous testing, while in 19 situations level of function was listed or described in psychiatric or other clinical reports. For the 19 individuals with no IQ scores available, reliability of cognitive

functioning rating was checked by an independent evaluator. The evaluator reviewed one-third (7) of these records, and a Pearson product-moment correlation was used to determine the degree of agreement between ratings. Use of a Pearson product-moment correlation is recommended by Hartmann (1977) for calculating reliability for continuous variables. Results showed satisfactory reliability with a correlation of .87 (2-tailed; p=.012) between the two ratings.

In the case of one participant, no IQ information or descriptive clinical reports were available, and it was necessary to use caregiver description to decide on level of functioning. For this individual, the level of functioning was clearly very high since the person lived independently in the community and held a competitive job. Assignment in this case was based on consultation with caregivers and clinicians involved with the individual.

Dependent measures. The scores on the DSDS (Gedye, 1995) were used to assess decline in all participants. The DSDS is a caregiver-based interview which yields a numeric score corresponding to the number of symptoms present, and therefore the severity of dementia. The DSDS also takes into account duration of symptoms and suggests a cut-off score for the diagnosis of dementia. The American Association on Mental Retardation and the International Association for the Scientific Study of Intellectual Disability both recommend the DSDS as an appropriate and comprehensive instrument for the assessment of dementia in adults with Down syndrome (Aylward, et al., 1997). The recommendation of this scale by the aforementioned associations was based on a review of a variety of currently available instruments constructed for adults with Down syndrome (Aylward et al., 1997).

The reliability of scores on the DSDS was examined by obtaining

ratings from two independent clinicians for 11 participants, or one-third of the sample. Level of agreement between scores was calculated using a Pearson product-moment correlation. Results of this investigation showed a relatively low reliability ($r=.69$, $p<.018$). It should be noted, however, that since AD is a progressive disease, many clients continued to deteriorate during the time between the two ratings, and this was reflected in higher deterioration scores at the time of the second rating. While ratings were done as closely together as possible, in some cases there was up to six months between each rating. As a result, reliability between ratings may be lower by virtue of this time lag in collecting the second set of data.

Each participant also received a diagnosis from the neuropsychologist. Based on psychometric testing and clinical interview, each participant was judged by the neuropsychologist to fall into one of three categories; no dementia present, possible dementia, or probable dementia. A diagnosis of no dementia was given when no symptoms were present. A diagnosis of possible AD was given when symptoms of dementia were present, but it was not possible to rule out all causes other than AD. A diagnosis of probable AD was given when symptoms of dementia were present, and most or all other causes had been eliminated from consideration (Aylward, Burt, Thorpe, Lai, & Dalton, 1995).

Results

Independent Variables

Information regarding the age and sex of all participants is presented in Table 1. Age was approximately normally distributed in this sample, and the split of male to female participants was 61% to 39%, respectively. Information regarding level of cognitive functioning for all participants is presented in Table 2. Level of functioning was also approximately normally distributed. Scores on the Residential Lifestyles Inventory were relatively normally distributed, and are presented in Table 3. Residential Lifestyles Inventory scores were unavailable for three of the 36 participants so analysis of this variable is based on 33 cases only. Data regarding years of schooling was divided into four categories, or levels, and is presented in Table 4. The resulting variable, level of schooling, was approximately normally distributed. Level of schooling was unavailable for four participants, therefore analyses are based on 32 cases.

Data regarding the number of years spent in an institution by participants is presented in Table 5. A large number of individuals did not spend any years in an institution so the curve for this variable had a rather severe negative skew. Transformations failed to improve the distribution of this variable, therefore it was left in its original state. Information regarding type of employment was collapsed into six categories. Employment setting ranged from day programs, to sheltered workshops to competitive employment environments. Hours of employment varied from part-time to full-time. Table 6 presents a description of the categories used for analysis and the number of participants in each category. The variable employment was relatively normally distributed. Table 7 shows probability values regarding skewness and kurtosis for each variable. Probability values reflect whether

Table 1

General Demographics of Participants

Variable	Frequency	% of Total
Sex		
Male	22	61
Female	14	39
Age (in years)		
29-35	6	17
36-45	10	28
46-55	14	39
56-65	5	14
65+	1	2

Table 2

Number of Participants at each Level of Cognitive Functioning

Level of Delay	Frequency	Percent of Total
Mild	3	8.0
Mild-Moderate	7	19.5
Moderate	11	31.0
Moderate-Severe	8	22.0
Severe-Profound	7	19.5

Note: "Level of Delay" categories are based on DSM-IV classification criteria.

Table 3

Distribution of Participants' Scores on The Residential Lifestyles Inventory

Range of Score	Frequency	% of Total
0-10	1	3
11-21	5	15
22-32	13	39
33-43	11	33
44-54	2	7
55-65	1	3

Note: Information was available for only 33 participants.

Table 4

Level of Schooling Completed by Participants

Level of Schooling	Frequency	% of Total
None	4	13
Finished Public School	2	6
Finished Secondary School	20	62
Additional Schooling (e.g. Adult education programs)	6	19

Note: "Additional Schooling" refers to participants who finished secondary school and continued in some type of educational program past the age of 21 years old.

Information regarding schooling was available for only 32 participants

Table 5

Number of Years Spent in an Institution by Participants

Range of years	Frequency	% of Total
None	25	69.0
Between 5 and 7 years	3	8.0
Between 15 and 20 years	4	11.5
More than 20 years	4	11.5

Table 6

Employment Characteristics of Participants

Employment Variable	Frequency	% of Sample
Category of Work		
No employment/day program	3	8
Part-time in a sheltered workshop	6	17
Part-time with some competitive jobs	5	14
Full-time in a sheltered workshop	16	44
Full time with some competitive jobs	4	11
Full-time with mostly competitive jobs	2	6

Table 7

Skewness and Kurtosis of Each Variable

Variable	Skewness p*	Kurtosis p
Age	.39	.17
Level Functioning	.13	.38
Yrs. in Institution	.00	.00
RLI**	.44	.21
Yrs. Schooling	.008	.16
Employment	.23	.33
DSDS (raw)***	.08	.00
DSDS (categorized)	.07	.42

Note: *p means the probability that either the skewness or the kurtosis is significantly different from normal. Probabilities of less than .01 represent a significant departure from normality.

RLI** means Residential Lifestyles Inventory.

DSDS*** means Dementia Scale for Down Syndrome.

the skewness and kurtosis of a distribution are significantly different from normal.

Upon inspection of the data it was found that very few participants smoked cigarettes or took anti-inflammatory drugs. Because of the small numbers, it was impossible to analyse this data. Summary information regarding these two variables is included in Table 8, however the variables were not included in the final analysis.

Dependent Variable

As it was impossible to obtain DSDS scores for the "nursing home participant" and the two deceased participants, the mean value of the group of all demented subjects was used for these three participants. This constitutes a conservative estimate of their scores since the two deceased individuals were in the late stages of dementia when they died, and the nursing home participant is currently in the late stages of AD. This means their scores would have likely been substantially higher than the mean of those individuals seen for assessment, who were all in the early to middle stages of the disease.

Raw scores on the DSDS ranged from 0, indicating no symptoms, to 23 indicating many symptoms. The distribution of DSDS scores had a significant negative skew (see Figure 1). In order to normalize the scores, a number of transformations were attempted but none was found to produce an acceptable solution. As a result, raw DSDS scores were placed into six composite categories, a procedure which was found to render the new distribution less skewed. This new DSDS category score replaced the raw DSDS score and was used in all analyses as the dependent variable. The distribution of the DSDS composite categories is shown in Figure 2. A description of the categories is shown in Table 9.

Table 8

Smoking and NSAIDs Information for Participants

Characteristic	Frequency	% of Sample
Used NSAIDs	8	22
Smoked Cigarettes	2	5
Exposed to Cigarette smoke	5	14

Note: "NSAIDs" refers to non-steroidal anti-inflammatory drugs. Amount of this substance used by participants varied greatly. "Exposed to Cigarette smoke" is defined as someone who lived in the same home as the participant smoked cigarettes.

Reliability data gathered on the DSDS was recalculated using the category system described above, and a Pearson product-moment correlation was applied to these data. Results indicated an improved reliability of scores ($r=.79$, $p=.004$) from that previously obtained ($r=.69$, $p=.018$).

Subsuming the raw DSDS scores into six categories was also found to be useful for purposes of combining the quantitative rating on the DSDS questionnaire and the neuropsychologist's ratings. Under the new scheme, a score of 1 corresponded to a DSDS score of 0, or no symptoms of dementia present. A score of 2 corresponded to a few minor symptoms being present, which by themselves were deemed insufficient to diagnose dementia. A score of 3 corresponded to the presence of four to nine symptoms on the DSDS, and a diagnosis of possible dementia; a score of 4 meant multiple symptoms were present and a diagnosis of possible dementia in the early stages was given. A score of 5 meant between 16 and 21 symptoms were found to be present and a diagnosis of probable dementia was given, while a score of 6 meant a diagnosis of probable dementia with 22 or more symptoms present. Table 9 gives a summary of the correspondence between neuropsychologist rating and DSDS scores. The numbers shown for each category in Table 9 were used for all analyses.

Analyses

Prior to the analyses, all variables were examined through SPSS for Windows (Version 6.1) for accuracy of data entry, missing values, normality, homoscedasticty and all the assumptions of the multiple regression model. As well, an examination of univariate and multivariate outliers was undertaken. Results of these investigations showed that variables met the assumptions of multiple regression and no outliers were found.

The intercorrelation matrix among variables is presented in Table 10.

Table 9

Dementia Scale for Down Syndrome Composite Scores: Clinician Ratings Combined with Dementia Scale for Down Syndrome Scores

Category	Clinical Rating	DSDS symptom score range
1	no dementia	score of 0 (no symptoms)
2	no dementia	score of 1-3 (a few minor symptoms)
3	possible dementia	score of 4-9 (sub-clinical dementia)
4	possible dementia	score of 10-15 (early stage dementia)
5	probable dementia	score of 16-21 (early to middle stage)
6	probable dementia	score of over 22 (middle stage dementia)

Table 10

The Intercorrelation Matrix for All Variables

Variable	2	3	4	5	6	7	8
1. Age	.133	-.253	-.378*	.353*	-.447**	-.212	.527**
2. Sex	--	-.212	-.621**	-.042	-.133	-.070	.144
3. Level of function		--	.566**	-.364*	.457**	.218	-.484**
4. Level of School			--	-.131	.340	.121	-.169
5. Years in institution				--	-.252	.051	.201
6. Employment					--	.281	-.238
7. RLI[1]						--	-.110
8. DSDS score							--

Note: * $p<.05$, two-tailed. ** $p<.01$, two-tailed.

RLI[1] means Residential Lifestyles Inventory.

For the variable "Sex", male participants were coded as 1 and female participants as 2.

As can be seen, there were a number of relatively high correlations between the independent variables, as well as between the independent variables and the dependent variable. For example, the correlation between level of schooling and level of functioning was .56 ($p<.001$, 1-tailed), indicating that higher level of functioning was associated with more years of schooling. The correlation between level of functioning and employment was .45 ($p<.005$, 1-tailed), indicating that more challenging employment was also associated with higher levels of functioning. Finally, the correlation between level of functioning and years in an institution was -.36 ($p<.03$, 1-tailed), suggesting that individuals with lower levels of functioning have spent more time in institutions.

The first analysis of data was accomplished through the use of multiple regression. Although Tabachnick and Fidell (1989) suggest that the minimum ratio of independent variables to subjects in multiple regression analysis should be at least one to five, Howell (1997) argues that a ratio of one to ten is more appropriate. In the present study, if all independent variables were entered into a single equation, the ratio of predictors to subjects would have been one to six. Because this is at the low end of the acceptable range, steps were taken to decrease the number of predictors entered and it was decided that the analysis would be completed in three stages. In the first stage, age, level of functioning, years of school, years in an institution, employment score and RLI score were all entered into a regression equation with the DSDS composite score as the dependent variable. A stepwise entry procedure was used due to the large amount of shared variance between independent variables, as shown in the intercorrelation matrix. By using a stepwise entry, it was expected that the best predictors would be identified. Using this method, age entered the equation first, followed by level of

functioning, F (2, 27)=8.32, p=.001. Both age (p=.008) and level of functioning (p=.02) were significantly related to DSDS composite. No other variables entered the equation. Note that the degrees of freedom were reduced in this analysis due to missing data points (see Tables 3 and 4).

In the second step, the two variables found to be related to DSDS composite in the first equation, namely age and level of functioning, were entered into a new regression equation by themselves. Using a stepwise entry procedure, it was found that age entered the regression equation first, followed by level of functioning. Results of the analysis indicated that together, age and level of functioning significantly predicted DSDS composite, F (2,33)=11.45, p=.002. As expected, results showed that older participants were more likely to develop symptoms of dementia (p=.003). Level of functioning was also significantly related to DSDS composite (p=.01). As predicted, those with a lower level of functioning were more likely to develop symptoms of dementia. Semi-partial correlations revealed that age accounted for 17.5% of the variation in DSDS composite, while level of functioning accounted for 13%. Each semi-partial correlation describes the variation that is unique to either age or level of functioning after the variation associated with the other is removed (Tabachnick & Fidell, 1989). Thus, a total of 30.5% of the variation in DSDS composite is accounted for by these two variables.

The third step of the analysis was undertaken to investigate the contribution of the environmental stimulation variables when they were all forced into the regression equation, regardless to their significance. Semi-partial correlations were calculated for each variable to discover the unique variation contributed to the equation by them. Results of this analysis showed that taken together, years in an institution, employment, and RLI score contributed only 2.6% to the total variance in DSDS composite scores. Years of schooling

contributed 8% to the total variation, but it was in the opposite direction to that expected, with more years of school predicting higher scores on the DSDS composite. This opposite to expected relationship only emerged after the effect of level of cognitive functioning was removed. Examination of the intercorrelation matrix shows that, on its own, years of schooling is negatively related to DSDS score, meaning that more years of schooling predicted lower DSDS composite scores.

Although collapsing DSDS scores and clinician diagnosis into DSDS composite scores was useful for interpretation and helpful in meeting the assumptions of multiple regression, it is interesting to note that in a previous analysis using uncategorized (raw) DSDS scores, results similar to those described above were obtained. Again, age and level of functioning were found to significantly predict symptoms of decline, but with slightly different probability values. This suggests that categorizing DSDS scores did not alter their relationship to the independent variables.

Post-Hoc Analysis. The relatively high correlations between level of functioning and years of school, employment, and years in an institution suggested that the relationship between these variables required further exploration. For this reason a post-hoc multiple regression was performed using years of school, years in an institution and employment as the independent variables and level of functioning as the dependent variable. This was done to explore whether years of school, years in an institution and employment are better understood as relating to level of functioning, rather than DSDS composite. Results showed that years of school, employment, and years in an institution significantly predicted level of functioning, $F(3, 28)=7.41$, $p=.008$. Semi-partial correlations show that years of school, employment, and years in an institution predicted 20%, 6%, and 5% of the variation in level of functioning, respectively.

Discussion

The aim of this study was to investigate possible protective factors for the development of AD in adults with Down syndrome. Results suggest that higher functioning adults with Down syndrome are less likely to have decline consistent with AD than are their lower functioning agemates. If higher level of cognitive functioning can be interpreted to reflect greater synaptic density, or higher neuronal reserve, this would seem to support the hypothesis that increased neuronal reserve may defer the clinical expression of AD. However, variables measuring participant activity and level of stimulation in the environment were not found to be significantly associated with the onset of decline in this study. Since greater environmental stimulation was expected to be associated with greater neuronal reserve, this is contrary to the predictions of the neuronal reserve hypothesis. It is possible, of course, that were a larger sample to have been examined, these factors might have been shown to be relevant.

As expected, age predicted AD symptom onset. It is well established that advancing age is one of the most reliable predictors of AD in all populations (e.g. Holland, 1995; Mann, 1988; Zigman, Silverman, Wisniewski, 1996). The present findings therefore correspond well with established information in this area.

Although few researchers have examined the matter specifically, studies of adults with Down syndrome have not generally found level of functioning to be predictive of AD (Evenhuis, 1990; Lai & Williams, 1989; Visser, Aldenkamp, Overweg, & Wijk, 1997). The reason for this may be related to restricted sample selection in previous studies. As Zigman (1993) noted in his review of the literature concerning Down syndrome and AD,

most previous research in this area has included only those who lived in institutions. This is because it is much simpler, from a methodological point of view, to study individuals all living in one place and because in the past, many more individuals with Down syndrome resided in institutions. However, research which included only those in institutions likely suffered from a restricted range in terms of the cognitive ability of the participants included, since institutional samples generally have a preponderance of moderately and severely delayed individuals. This may have made any differences in the rates of dementia between groups more difficult to detect.

A number of more recent papers have attempted to gather information on community-based rather than institutional samples. An interesting example is the study by Devenny et al. (1996). These researchers reported the results of a five-year study of Down syndrome individuals, aged 31 to 63 years, with mild to moderate delay, who were living in the community. Over the period of the study, only four of their 91 participants (4%) showed any symptoms of decline, and no difference in rates between high functioning and moderately functioning individuals was reported. Although it included more high functioning individuals, again the range of ability in the sample was restricted since no lower functioning individuals were included.

The overall rate of diagnosis found in the Devenny et al. (1996) study could be interpreted to lend support to the findings in the present study. The 4% rate of dementia reported by these authors is in sharp contrast to the 51% reported by Lai & Williams (1989) in their predominately institutionalized sample. Although the methodologies differed somewhat between these two studies, taken together their results would seem to support the notion that higher functioning individuals may experience a deferral of the symptoms of AD. As well, it may be the case that community living offers a more

stimulating environment and this also works to decrease the incidence of dementia (Devenny et al., 1996).

Investigation of the variable "level of cognitive functioning" showed that it was highly correlated with a number of environmental factors. Results of a post-hoc multiple regression showed that higher level of functioning was associated with more years of schooling, more challenging employment, and fewer years in an institution. These results suggest that the relationship among these variables and symptoms of dementia may be more complex, and more indirect, than expected. Since level of functioning is significantly related to symptoms of dementia, while years of schooling, years in an institution, and employment are significantly related to level of functioning, the relationship of the environmental enrichment factors to symptoms of dementia may be mediated by level of functioning. In other words, it may be that years of school, years in an institution and employment influence level of functioning, and level of functioning, in turn, influences symptoms of dementia. Figure 4 shows a diagram of how this proposed relationship between variables might operate.

Although this study did not address the issue of the relative contribution of genetic versus environmental factors to the development of level of cognitive functioning, the question is nonetheless of relevance to this study's results. The magnitude of the correlations between the variables discussed above suggests a relatively large degree of shared variance between level of functioning and the environmental variables. In fact, together years of school, years in an institution and employment account for 31% of the variation in level of functioning. Because of this shared variance, it is difficult to know whether level of functioning, as it is defined here, measures innate factors, environmental factors, or some combination of both. It could

be that biologically determined cognitive ability influences such things as length of school exposure and employment experiences. Conversely, it could be that having particular environmental experiences, such as many years of schooling or a challenging job, influences an individual's level of cognitive functioning. It is also possible that both explanations are correct.

In the context of this study, level of functioning can be seen as an approximation of full scale IQ. As a result, all of the varied arguments which can be mounted regarding the origins of variation in IQ may be applied. Proponents of the genetic explanation might point out that a substantial portion of IQ is accounted for by inheritance (e.g. Bouchard, Lykken, McGue, Segal & Tellegen, 1990; Scarr, 1993). On the other hand, proponents of an environmental model would argue that there are equally important environmental factors at work modifying genetic factors (Baumrind, 1993; Neisser, et al., 1996; Rutter, et. al., 1997).

Neisser et al. (1996) offer a theory regarding IQ which seems especially helpful for addressing the question of genetic and environmental contributions. These authors point out that in children, genetic factors account for approximately 40% of the total variance in IQ. However, as people become older, the variance accounted for by genetics increases. By adolescence, it grows to approximately 75% . Neisser et al. (1996) explain this change by suggesting that as children grow, they are increasingly able to select their own environments. These selections may create a kind of genotype-environment interaction, which serves to strengthen the existing genetic predisposition. An explanation such as this argues for a strong genetic component to IQ, but it also speaks to the malleability of IQ through environmental interactions.

Applying Neisser et al.'s (1996) theory to the outcome from this study

would suggest that level of functioning, as it is described here, might be best characterized as a composite variable reflecting the outcome of a variety of innate and environmental factors. An implication of this may be that interventions designed to enrich the environment of adults with Down syndrome may be effective in increasing their level of cognitive functioning, and perhaps concurrently deferring the symptoms of AD. By providing a wider and more enriched environment, it may be possible to promote greater ability since having a greater selection of activities would presumably allow individuals with Down syndrome to make their selections based on genetic predisposition. This may result in more active and motivated choices (Neisser et al., 1996).

Having said that level of cognitive functioning maybe a composite of environmental and innate factors, it must be acknowledged that all environmental factors examined in this study failed to demonstrate a significant relationship to symptom score. There are a number of possible reasons for this failure. Perhaps the most obvious of these is the small sample size studied here. Given a larger group other factors might have emerged as predictive. Partial correlations showed that the environmental variables accounted for a very small unique portion of the total variance, however, even in combination, their effect was slight. It is also possible that some important environmental variable was missing from this equation. For example, it may be that number of friends or social contacts a person has influences the amount of environmental stimulation they receive. As this and many other variables were not studied, their effect remains unknown.

It is also possible that genetic or congenital factors account for most of the variance in level of functioning in adults with Down syndrome, and the effect of the environment is minimal. Neisser et al.'s (1996) theory is based

on data from the general population, and those with mental retardation may differ from the general population in some important way that we have not as yet recognize. However, even if environmental factors are of minimal importance and genetics play the most important role, it is still of use to establish which individuals are at the greatest risk for AD. With the recent approval of the drug Donepezil to treat AD, early identification of the disease has become an important priority for research endeavours (Bryson & Benfield, 1997).

Although the information from this study has been interpreted to support the relatively broadly defined neuronal reserve hypothesis, other explanations for the relationship between symptoms of decline and level of cognitive functioning could also be suggested. For example, it could be that individuals with a lower level of functioning experience the ill effects of aging earlier than those with a higher level of functioning. This argument is supported by the fact that individuals with a severe to profound delay die earlier relative to those with a milder delay (Eyman, Call & White, 1989). In effect, at the same chronological age, lower functioning individuals are actually at a later period in their lives. It might be supposed that earlier death is accompanied by the earlier onset of health and aging problems which may mimic dementia. This could certainly account for symptoms such as slowed motor responses or poorer memory ability. However, it may not adequately explain other AD symptoms such as irritability or mood swings. In any case, it is possible that at least some of the variation in symptoms of decline found in this study may be attributable to differential aging effects.

Study Limitations

A number of limitations, which may have influenced the results in this study, must be acknowledged. First, the size of the sample examined here

is quite small, a factor which reduces the power of the analyses for detecting differences between groups. Thus, although no significant effects were found for the environmental variables, their influence cannot be ruled out on the basis of this data. This is especially true for the variables "years spent in an institution", "use of anti-inflammatories" and "cigarette smoking", since very few participants fell into the exposure groups for these variables. Larger samples and better controlled studies in this area will be necessary for studying these variables in the future.

A second problem which must be acknowledged is that of degree of diagnostic certainty. Many of the participants in this study were relatively recent additions to Surrey Place's client group. For this reason, it was not possible to follow their progress over an extended period of time. Six months is the minimum duration of symptoms necessary to diagnose AD, and although all clients were followed by the clinic for at least this long, in many cases all other potential reasons for decline could not be ruled out within this time period. Extended observation, as well as post mortem examination, would be necessary to rule out other causes of decline, such as multi-infarct dementia.

One especially difficult issue in the area of differential diagnosis is the separation of decline associated with depression from decline associated with AD. In developmentally delayed adults, depression is often manifest behaviourally as apathy, loss of skills, irritability, and/or sleep disturbance (Pary, 1992). These symptoms are also some of the first signs of decline in AD. Depression is relatively common among adults with developmental delays, and studies have found rates of depression to vary from 10% to over 40% for this group (Gilley, 1993). As well, depression has been documented to co-exist with AD making the separation of symptoms even more difficult (Pary, 1992).

As advanced maternal age is a risk factor for having a Down syndrome child, at the age of 40 an adult with Down syndrome is likely to have parents over the age of 80 who may be nearing the end of their lives. As a result, when individuals with Down syndrome are facing the greatest risk for AD, they are also often dealing with the death of their parents or other family members. This possible co-ocurrance of AD symptoms and depression makes differential diagnosis a very complex task.

Finally, the issue of detection of dementia symptoms in individuals of various ability levels is a constant difficulty. As discussed earlier, the first symptoms of dementia are often more obvious in those with higher levels of functioning since they engage in more complex activities (Aylward, Burt, Thorpe, Lai & Dalton, 1997). The lose of higher abilities, such as reading or writing skills, is also much easier to detect with psychometric testing procedures than are changes or loses in other areas, so the diagnosis of individuals with higher abilities can often be accomplished with less difficulty (Shepperdson, 1995). This is likely the reason why, in this study, after the effect of level of cognitive functioning was removed, individuals with more years of schooling appeared to display more symptoms of decline. It should be noted, however, that the fact that level of functioning remained a significant predictor in the face of this difficulty makes the results from this study all the more compelling.

Applied Implications of these Findings

Research into factors that may influence the onset of dementia in the Down syndrome population has implication for both prevention and intervention. In the realm of prevention, support for the existence of protective factors found through studies such as this may enable health care professionals to take steps to reduce or delay the onset of dementia in future

generations of Down syndrome adults. Deferral of symptom onset for even a short period of time would result in substantial savings in the cost of treatment and care for patients, not to mention the emotional costs to victims, their caregivers, and their families.

In the field of intervention, details regarding which factors are responsible for an increased risk of dementia could aid in the early identification of symptoms in individuals with Down syndrome. Early identification will help to ensure that appropriate supports and management plans can be established to help the patient's family adjust, and to provide increased assistance to the patients themselves. Early counseling and education regarding AD will aid caregivers in coping with the increasing levels of disability inevitable with AD, and assist them in identifying and utilizing available sources of support. As well, with the recent approval by the Canadian government of the drug Donepezil (Bryson & Benfield, 1997) for the treatment of AD, early diagnosis will help in providing the appropriate medication to those who can benefit from it, as quickly as possible.

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Appendix 1

Information Letter For Caregivers
Project: Examining the Aging Process in Down Syndrome

Surrey Place Centre and the University of Guelph are working together to study the aging process in Down syndrome. We are asking for adults with Down syndrome and their caregivers to help us by participating in a research study.

The purpose of the study is to examine how the abilities of Down syndrome adults change as they get older. We wish to investigate factors which may protect against the loss of abilities or memory with aging. As you may know, adults with Down syndrome are at increased risk for developing memory problems and adaptive behaviour problems as they age, so it is important for us to examine factors which may decrease this risk.

The study involves three parts. First, you will be asked some questions about (client's name). The questions concern (client's name)'s schooling, hobbies and past and present jobs. We will also ask about his/her health, ability to care for him/herself, history of illnesses, and some personal habits such as alcohol consumption or smoking habits.

The second part of the study involves reviewing (client's name)'s records at Surrey Place Centre to find out about past events in his/her life. We will look for additional information about health, jobs, schooling and past activities.

The third part of the study involves comparing (client's name)'s neuropsychological test scores at this visit to past scores to see if there has been any change.

All of the information collected in this study will remain completely confidential and will be stored safely. Identifying information will only be available to myself (the Examiner), Dr. Emoke Joszvai (the Project Director) and Dr. Mary Konstantareas (University of Guelph supervisor).

As is always the case at Surrey Place Centre, a follow-up interview will be conducted to explain test results and provide or suggest additional services as necessary.

Participation in this study is completely voluntary. If you chose not to participate, it will in no way effect your access to the many services offered at Surrey Place Centre.

We hope you will be able to help us, as your participation is very important.

Information letter for Clients

We are doing a study and we would like you to help us. We want to know about how people with Down syndrome change as they get older.

The study has three parts. First, we will ask your caregiver some questions about you. We will ask about where you went to school, what you like to do and where you have worked. We will also ask about your health.

Next, we will look at your records at Surrey Place to find out more about you.

Then, we will look at your test scores from before and from today to see if they have changed.

After this we will meet again to talk about your tests and the results.

You don't have to be in this study if you don't want to. Everything will stay the same for you at Surrey Place if you decide not to be in the study.

We hope you will help us because your help is very important.

Thank you

Appendix 2

Consent Form for Clients

I, _____ know that I am going to do some
(client's name here)

tests at Surrey Place. I know this student will look at the tests and use them in a study.

I know this student will ask my caregiver some questions about me. The questions are to help people understand Down syndrome better. She will also read about me in my file at Surrey Place Centre.

I can say NO if I don't want to do this.

I have read this. I will help with this study.

Signed:_____ Date: _____

Witness: _____ Date: _____

Consent Form for Caregivers

I, _____ understand the general nature of
(caregiver's name here)

this study and the involvement required by my relative/client. I give

consent for _____ to participate in the study.
(client's name here)

I also agree to participate in the study myself.

I understand that participation in this study is completely voluntary, and refusal to participate in the study will in no way effect the quality of service I or my relative/client will receive from Surrey Place Centre.

I understand that I may refuse to answer any question, and may stop participating at anytime.

I understand that the investigators will review files held at Surrey Place Centre in order to gain historical information for this study.

I understand that the identifying information from our participation in this study will be kept strictly confidential at all times and will only be available to Dr. Emoke Joszvai, Dr. Mary Konstantareas and Valerie Temple B. Sc.

Signed: _____ Date:_____

Witness: _____ Date:_____

Appendix 3

Down Syndrome Health and Activities Questionnaire

Respondent Name: _____

1. What is your relationship to_____ :
(write the name of your relative/client here)

   mother or father ❑ brother or sister ❑ paid caregiver ❑

   other_____

2. How long have you known him/her? _____Years

3. Where does your relative/client live now?_____

4. Where else has he/she lived? (number of years he/she lived each place)

   with another relative _____(years) in a group home _____ (years)

   in an institution_____(years)

   in another place (specify)_____

   he/she has not lived anywhere else ❑ I don't know where he/she lived before ❑

5. Does he/she currently hold a job? Yes ❑ No ❑

   If yes, please describe current job(s) in the space below:

	Type of Job	Company	# of Years	Part /Full Time	Supervised /independent
1					
2					

6. Please describe all his/her previous jobs below

	Job Title	Company	# Yrs	Part/ Full Time	Supervised /Independent
1					
2					
3					
4					

No previous jobs ❑

7. At what age did he/she start school?_____age ❑ don't know

8. What kind of program did he/she attend at school? (check all that apply)

	# of yrs in program
❑ residential program	_____
❑ program in a special school	_____
❑ integrated program in regular school	_____
❑ fully integrated classes	_____
❑ withdrawal program (both regular and special classes)	_____
❑ don't know	

9. How many years of schooling did he/she complete? _____years

❑ I don't know

10. What are the names of the schools he/she attended?

1. _____ For how long ____ (years)
2. _____ For how long ____ (years)
3. _____ For how long ____ (years)

11. Has he/she ever suffered from any of the following?

	Yes	No	Don't know	Presently Medicated
thyroid condition	❑	❑	❑	❑
arthritis	❑	❑	❑	❑
psychiatric disorders	❑	❑	❑	❑
asthma	❑	❑	❑	❑
heart condition	❑	❑	❑	❑

Please list other psychiatric disorders or long term illnesses he/she has suffered:

_____

_____

12. Has he/she used any of the following medications regularly (regularly means at least once per week for several weeks):

	How often	For how long
Aspirin	_____	_____
Anacin	_____	_____
Bufferin	_____	_____
Excedrin	_____	_____
Motrin	_____	_____
Tylenol	_____	_____
Actiprofen	_____	_____
Indocid	_____	_____
Entrophen	_____	_____

Other headache or pain reliever	How often	For how long
_____	_____	_____

(please give name above)

13. Please list all the medications he/she is currently taking:

_____

_____

14. Has he/she ever had a head injury in which he/she lost consciousness?

❑ Yes ❑ No ❑ don't know

If yes, how long was he/she unconscious?

_____ minutes _____ hours _____ days ❑ don't know

How old was he/she when this head injury happened? _____ years

15. Has he/she ever engaged in head-banging (e.g. hitting his/her head against hard objects)?

❑ Yes ❑ No ❑ don't know

If yes, how often did this occur? _____

For how long a period did this behaviour occur? _____

16. Has he/she ever experienced seizures?

❑ Yes ❑ No ❑ don't know

If yes, how old was he/she when they first occurred? _____ years

How often do ? ❑ daily ❑ weekly ❑ monthly ❑ other_____

How long do they generally last? _____

17. Has he/she ever smoked cigarettes?

❑ Yes ❑ No ❑ don't know

If yes, on average how much does he/she smoke per week?

❑ less than 1 pack. ❑ one pack. ❑ more than 1 pack.

For how long has he/she smoked _____months or _____years

18. Does anyone in his/her home (place of residence) smoke ?

❑ Yes ❑ No ❑ don't know

If yes, how many people smoke in his/her home? _____

For how long has he/she been exposed to cigarette smoke? ___years

19. Does he/she drink alcohol (wine, beer, spirits) ?

❑ Yes ❑ No ❑ don't know

If yes, how many drinks per week on average? ___

For how long? _____years

Can you describe any situations where it appeared that your relative/client was acting differently that he/she used to? (e.g. loss of skills, memory; strange behaviour)

_____

_____

_____

_____

_____

Residential Lifestyles Inventory (Revised)

Activities (Media)	Never	Occasionally	Often
1. Watch T.V.	0	1	2
2. Listen to radio	0	1	2
3. Play records/CDS	0	1	2
4. Use cassette player	0	1	2
5. Watch slides/home movies	0	1	2
6. Use videos	0	1	2
7. Read/view books, newspapers Magazines	0	1	2
8. Listen to talking books	0	1	2
9. Use computer	0	1	2
Activities (exercise)			
10. Walk	0	1	2
11. Jog	0	1	2
12. Ride a bike	0	1	2
13. Ride an exercise bike	0	1	2
14. Participate in exercises/calisthenics	0	1	2
15. Participate in aerobics/jazzercize	0	1	2
16. Use trampoline/rebounder	0	1	2
17. Weight training	0	1	2
18. Participate in track and field	0	1	2
19. Attend gymnastics	0	1	2
20. Technical dance (ballet, folk etc)	0	1	2
21. Play catch (eg. Frisbee, ball)	0	1	2
22. Play softball/baseball	0	1	2
23. Play basketball	0	1	2
24. Play football	0	1	2
25. Play soccer	0	1	2
26. Play rugby	0	1	2
27. Play hockey	0	1	2
28. Play volleyball	0	1	2
29. Play racket sports	0	1	2
30. Play golf	0	1	2
31. Play badminton	0	1	2
32. Rollerskate/skateboard	0	1	2
33. Ice skate	0	1	2
34. Snowshoe	0	1	2
35. Cross country/downhill ski	0	1	2

36. Swim/dive	0	1	2
37. Waterski	0	1	2
38. Sail/raft/canoe	0	1	2
39. Hike/backpack	0	1	2
40. Camp	0	1	2
41. Horseback ride	0	1	2
42. Yoga	0	1	2
43. Judo/karate/martial arts	0	1	2
44. Special Olympics (other)	0	1	2

Activities (games/crafts)

45. Pinball/video games	0	1	2
46. Board games/card games	0	1	2
47. Pool/billiards	0	1	2
48. Ping-pong	0	1	2
49. Darts	0	1	2
50. Lawn games	0	1	2
51. Bowling	0	1	2
52. Shuffleboard	0	1	2
53. Puzzles	0	1	2
54. Needle craft (knit, embroidery)	0	1	2
55. Weaving basketry	0	1	2
56. Art classes/craft classes	0	1	2
57. Draw/paint/calligraphy	0	1	2
58. Take photos	0	1	2
59. Make scrapbooks/photo albums	0	1	2
60. Miscellaneous art projects	0	1	2
61. Manage collections (eg.Stamps)	0	1	2
62. Grow garden/plants	0	1	2
63. Bird watching	0	1	2
64. Fish/hunt	0	1	2
65. Fly kites	0	1	2
66. Play instrument/singing lessons	0	1	2
67. Woodwork/finishing	0	1	2
68. Car repairs	0	1	2

Activities (Events)

69. Attend church	0	1	2
70. Watch sports (live)	0	1	2
71. Go to movie	0	1	2
72. Attend plays/concerts	0	1	2
73. Attend club meetings	0	1	2

75. Attend parties/dances	0	1	2
76. Plan/give parties	0	1	2
77. Instructional classes	0	1	2
78. Attend shows (art, flower, science)	0	1	2
79. Go to museums	0	1	2
80. Use library	0	1	2
81. Go to park	0	1	2
82. Go to Science Centre, zoo .etc.	0	1	2
83. Attend festivals	0	1	2
84. Watch parades	0	1	2
85. Attend circus/rodeo	0	1	2
86. Go on scenic rides	0	1	2
87. Participate in scheduled outings	0	1	2
88. Use sauna, whirlpool	0	1	2

Activities (Visits)

89. Receive/write letters to friends Family	0	1	2
90. Receive/make phone calls to friends Family	0	1	2
91. Visit with friends, family	0	1	2
92. Community overnite activities	0	1	2
93. Community outings	0	1	2

Personal (Food)

94. Use fast food restaurants	0	1	2
95. Use sit-down bars/restaurants	0	1	2
96. Plan meals	0	1	2
97. Buy & store groceries	0	1	2
98. Prepare meals	0	1	2

Personal (Space & Belongings)

99. Shop for personal items/clothes	0	1	2
100. Wash, dry put-away clothes	0	1	2
101. Iron, mend clothes	0	1	2
102. Clean/straighten room	0	1	2
103. Make bed	0	1	2
104. Change bed	0	1	2
105. Clean bathroom	0	1	2
106. Clean kitchen (e.g. oven fridge)	0	1	2
107. Wash dishes	0	1	2
108. Floor care	0	1	2
109. Clean windows	0	1	2

110. Garbage/recycle	0	1	2
111. Do yard chores	0	1	2
112. Care for pet	0	1	2
113. Manage prosthetic devise	0	1	2

Misc.

114. Takes medication	0	1	2
115. Interests not mentioned	0	1	2
116. Budgets money	0	1	2
117. Banks	0	1	2
118. Pays bills	0	1	2

Figure 1. The hypothetical relationship between variables.

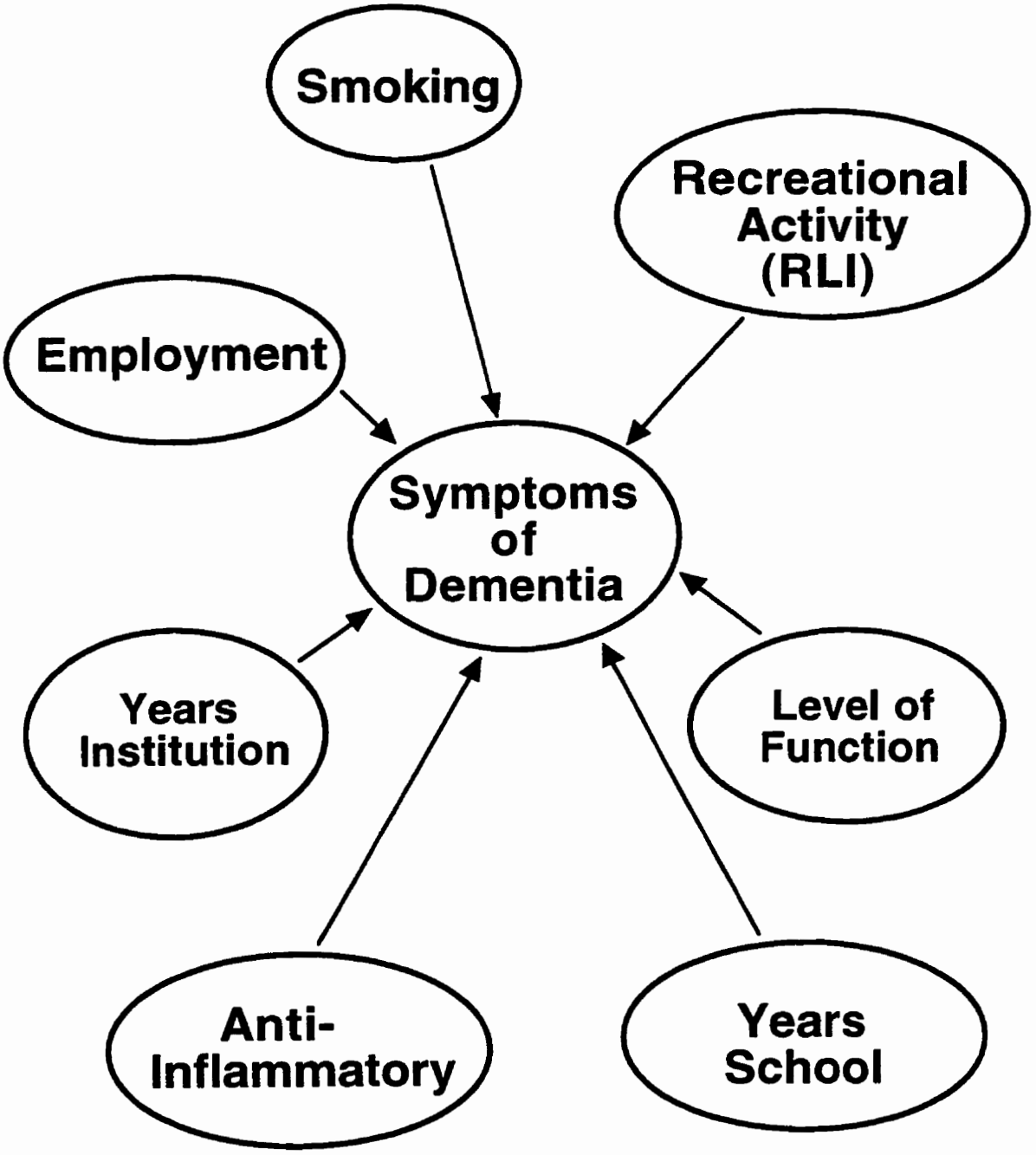
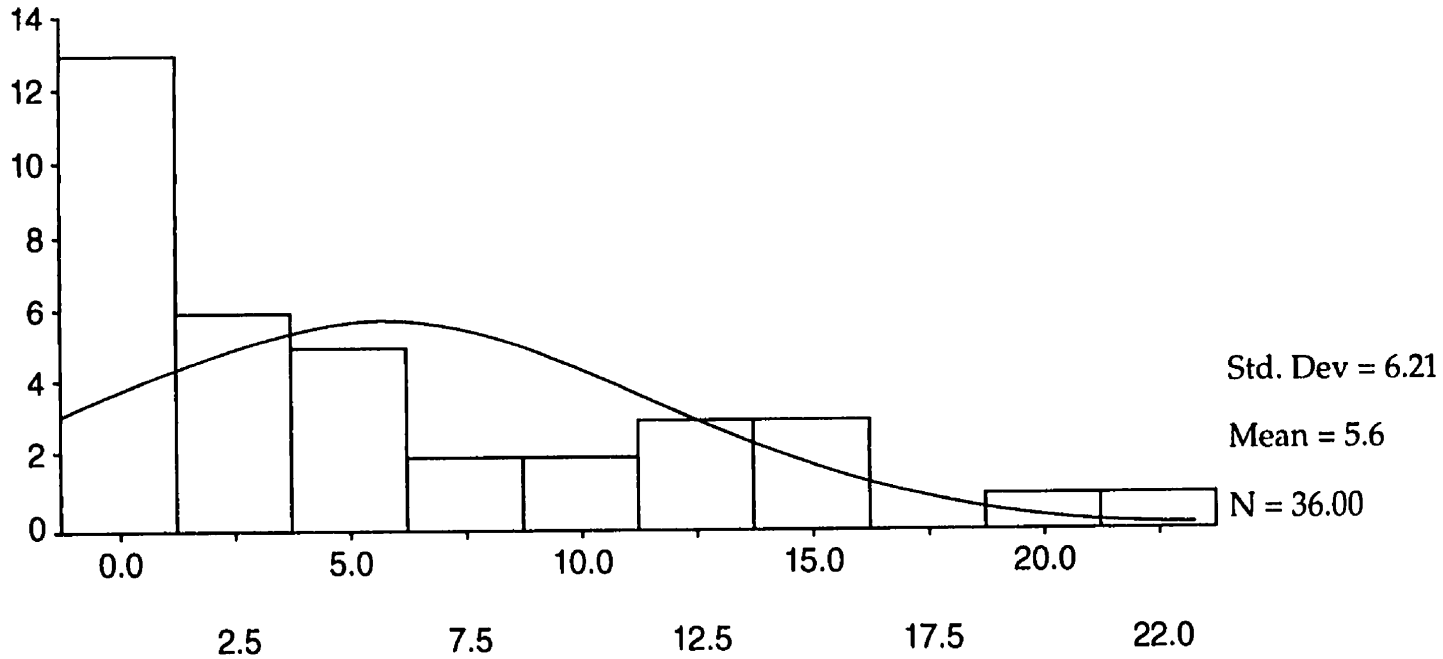


Figure 2. Raw Scores from the Dementia Scale For Down Syndrome



Raw Scores

Figure 3. Categorical Scores from the Dementia Scale For Down Syndrome

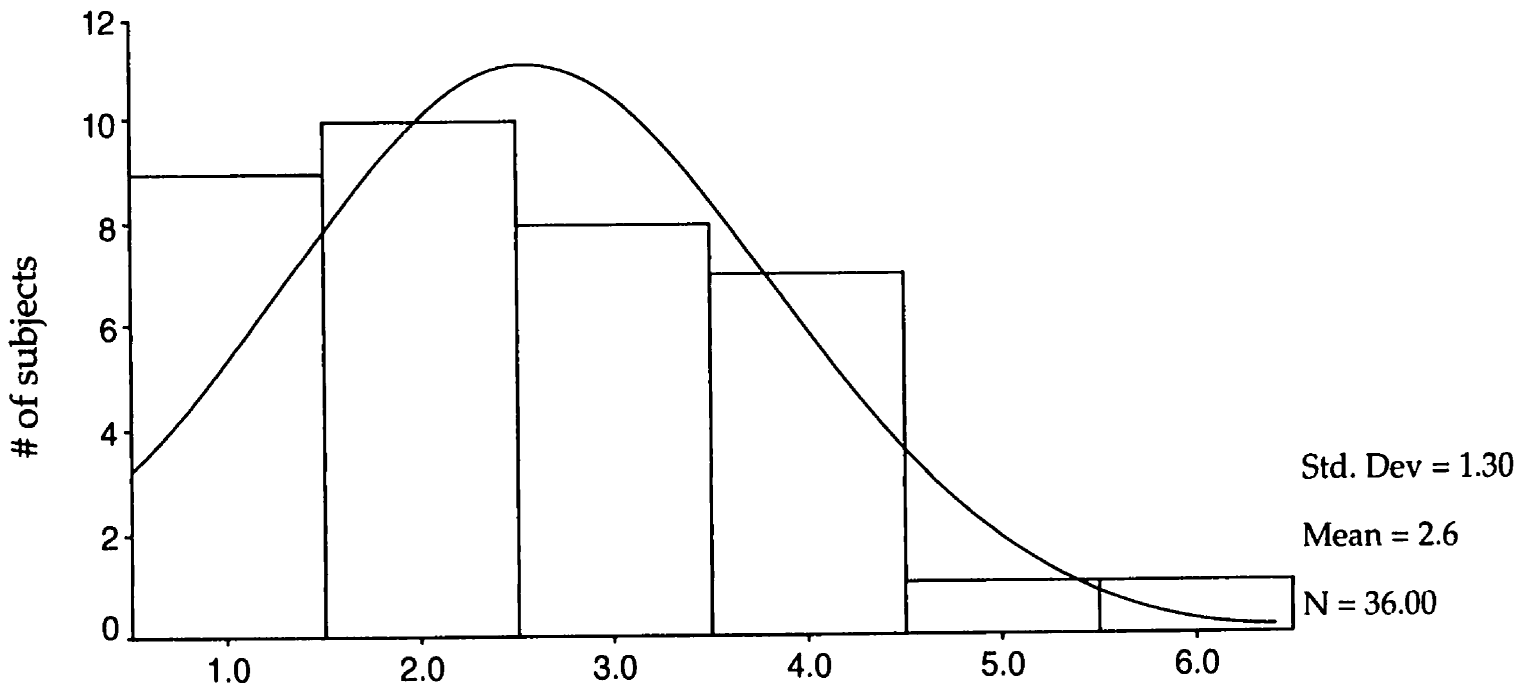


Figure 4. Relationship found between variables.

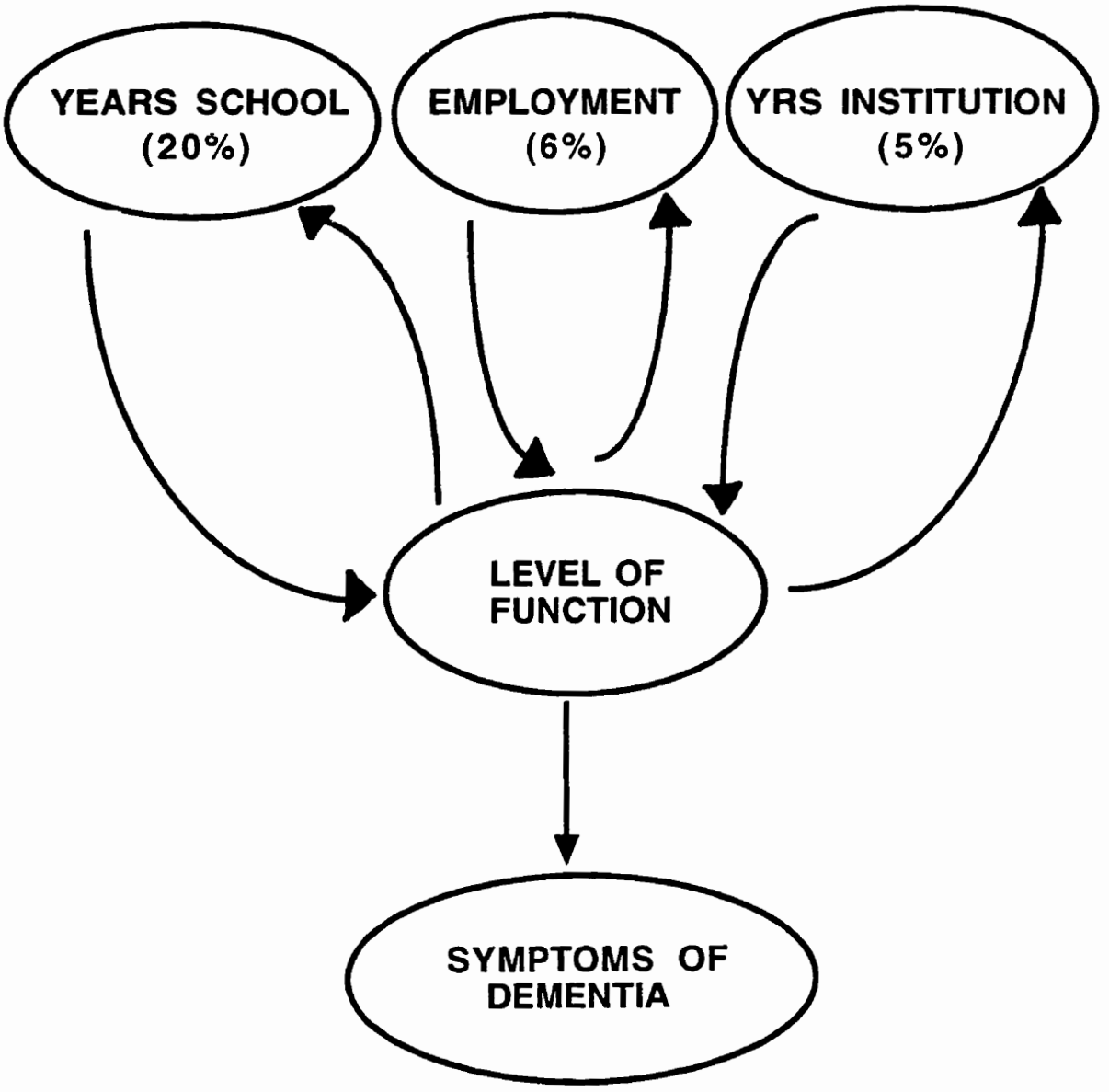
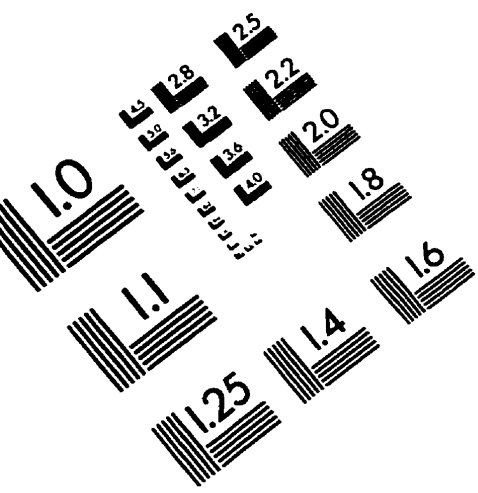
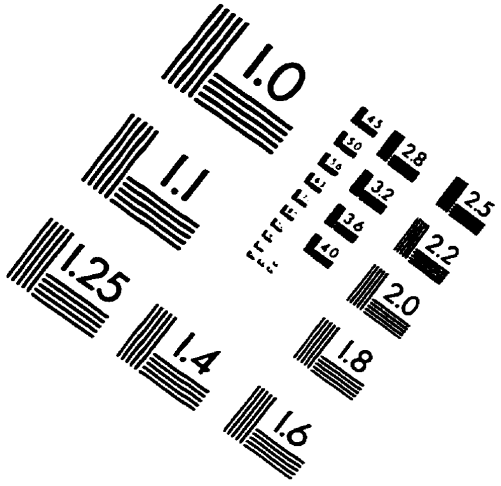
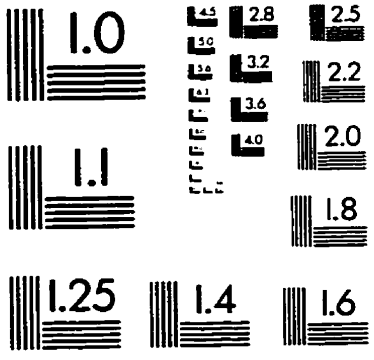
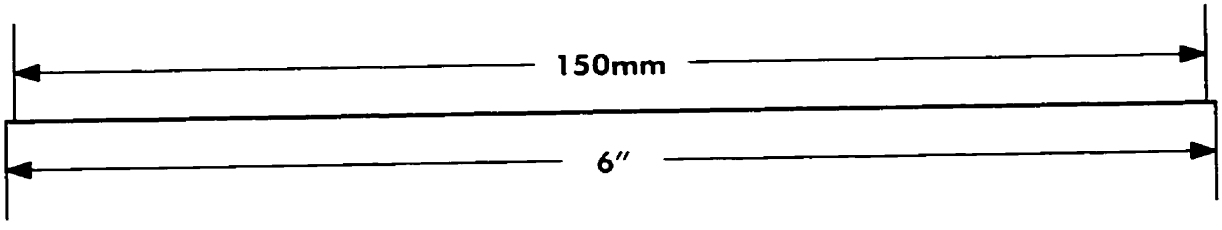


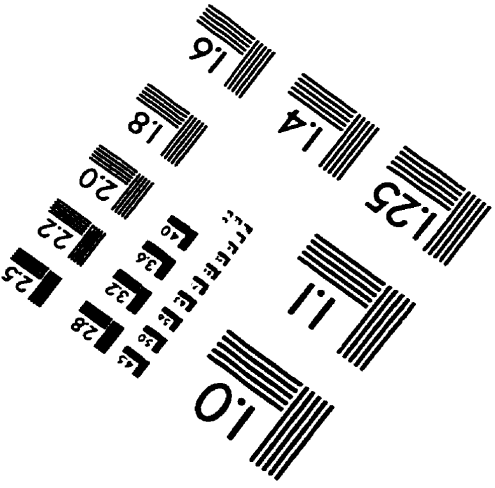
IMAGE EVALUATION
TEST TARGET (QA-3)

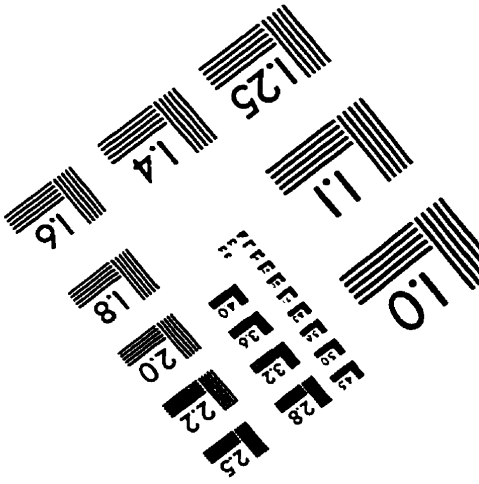












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