ADVANCING KNOWLEDGE INTO THE CLINICAL ASSESSMENT OF DEMENTIA

SIMON BENJAMIN NICHOLAS THOMPSON

A thesis submitted in partial fulfilment of the requirements of
Bournemouth University for the degree of
Doctor of Philosophy by Publication

September 2010

Bournemouth University

This copy of the thesis has been supplied on condition that anyone who consults it is understood to recognise that its copyright rests with its author and due acknowledgements must always be made of the use of any material contained in, or derived from, this thesis.

ADVANCING KNOWLEDGE INTO THE CLINICAL ASSESSMENT OF DEMENTIA

SIMON BENJAMIN NICHOLAS THOMPSON

ABSTRACT

The aim of the present thesis was to identify and measure cognitive and social abilities in people with dementia and learning disabilities.

In **Chapter 1**, normal ageing was discussed, distinguishing it from abnormal ageing and highlighting the problems it brings in terms of physical and psychological factors. Normal memory was also discussed and how the processes involved may be effected by dementia. Definitions of dementia were discussed in detail, outlining the neuropathology, neuropsychology and clinical signs of Alzheimer's disease, and the neuropsychology of multi-infarct dementia. The psychology of pseudodementia was presented and discussed and reference was made to the difficulties of a differential diagnosis. This was placed in the context of defining learning disabilities, such as Down's syndrome, and the

complexity of assessing people who have both a learning disability and dementia. Social and cultural differences were discussed together with the influence of environmental factors in measuring dementia. Theoretical considerations about the difficulties in assessing people with learning disabilities, particularly people who have Down's syndrome and dementia, were presented and discussed. These issues arose from the abnormalities in intellectual development and therefore, impacted upon subsequent cognitive rehabilitation and integration into the community of such individuals. A description and critique of the instruments used in the clinical studies ensued with important reference to the reliability, validity, and standard error and norms of each tool used. Finally, the goals of the study series were presented and discussed.

Part I: Empirical Studies (Published Papers)

Clinical studies were presented in chapters 2 – 5. All clients participating in the studies were randomly selected from consultant psychiatrists' lists of people living in their own homes or in voluntary sector group homes in England.

In **Chapter 2**, a neuropsychological test battery was devised in order to identify dementia in people with Down's syndrome. This battery comprised a series of standardised neuropsychological tests and rating scales measuring intellectual profiling, cognitive and social functioning and ability, and anxiety and depression. In addition to gathering biographical information, self-care information was also gathered. The Dementia Questionnaire for Mentally Retarded Persons proved to be a useful tool for indicating general areas of clients' skills that had declined; however, there is still a need for a definitive assessment of depression for these clients in order to discriminate between the effects of depression and those of dementia.

In **Chapter 3**, clients with Down's syndrome and non-Down's syndrome learning disabilities were assessed using specially selected neuropsychological assessment tools at two points separated by twelve months. Evidence was found to support hypothesis 1 which suggested that people with Down's syndrome show a greater decline in social abilities with age, compared with other groups of people with learning disabilities. Statistically, score changes reflecting the social abilities of the Down's

syndrome clients were found to be significantly greater ($p < .002$) than those of the non-Down's syndrome clients. Findings were explained in terms of poor language abilities in the Down's syndrome people generally, and the link between declining social abilities and dementia.

In **Chapter 4**, forty-one clients with learning disabilities were assessed using specially selected neuropsychological assessment tools at two time points separated by twelve months. Evidence for hypothesis 1 suggested that people with Down's syndrome show a greater decline in cognitive abilities with age, compared with other groups of people with learning disabilites. A weak linear association ($p < .004$; $r = .625$; 2-tailed) between cognitive performance and social abilities was also found for the Down's syndrome clients, supporting hypothesis 2. Findings were explained in terms of the link between declining cognitive abilities and dementia.

In **Chapter 5**, a clinical investigation was undertaken to determine the rate of decline in cognitive and social abilities in 16 clients with learning disabilities, 8 of which had Down's syndrome. Clients assessed at 6 months using specially selected neuropsychological tests and rating scales

measuring cognitive and social abilities as well as intellectual profiling. Both Down's syndrome and non-Down's syndrome clients were found to decline in cognitive abilities (Down's syndrome: $p < .005$; 1-tailed); Non-Down's syndrome: $p < .01$; 1-tailed; $p < .005$; 1-tailed). The Down's syndrome clients also showed decline in social abilities ($p < .005$; 1-tailed) over 6 months suggesting that changes between the two client groups may be significantly greater over a longer period, ie 12 months. Hence, the rate of change in cognitive abilities for the Down's syndrome clients was faster.

Part II: Evaluation of Treatment and Services (Published Papers)

In **Chapter 6**, a new version of the Benton Visual Retention Test for assessing people's memory functioning was evaluated. Findings showed that the conventional method of testing was preferred and not significantly different in terms of efficacy and reliability of measurement.

In **Chapter 7**, the potential benefits of Aricept, an acetylcholine esterase inhibitor, was investigated. There were significant effects and benefits for

patients who had mild-to-moderate Alzheimer's disease over a short period of time. However, the results were encouraging as they signalled the first documentation of the effect and a promising future for a possible remission of the disease.

In **Chapter 8**, a support group for wives of husbands with dementia was presented and discussed. The psychoeducation support group was shown to be an effective way of supporting newly diagnosed people with dementia and their carers.

In **Chapter 9**, a new interdisciplinary clinic for people with cognitive abilities was discussed. Importance in the constitution of the clinic personnel as well as the focus on assessment and follow-up treatment was emphasised.

Part III: Future Direction (Published Papers)

In **Chapter 10**, the importance of assessing people with dementia in the early stages of diagnosis and at particular intervals was demonstrated in

the context of the legal process. Suggestions were made for improving the test for testamentary capacity.

Part IV: In Summary

Discussion in **Chapter 11** covers the empirical work presented together with suggestions for future research, namely considering the differences between discrete types of dementias such as multi-infarct (vascular) dementia versus Alzheimer's disease and also longitudinal studies. Interesting findings from the clinical studies revealed a greater decline in social abilities of Down's syndrome clients versus non-Down's syndrome clients. These findings were explained in terms of poor language abilities in Down's syndrome clients generally, and the link between declining social abilities and dementia. A link between cognitive performance and social abilities was also found for the Down's syndrome clients. Findings of declining cognitive abilities in both groups of clients were associated with dementia; and in particular, with a failure of the Central Executive System and Articulatory Loop System, considered to be important in

normal memory. Modifying assessment techniques such as by computerisation is presented and treatment efficacy using the acetycholinesterase inhibitor, Aricept, is presented and discussed. The establishment of cognitive assessment clinics is also presented as a way of providing a comprehensive service for people with early onset dementia. Service implications for people with learning disabilities is discussed and finally, ways of improving the test for testamentary capacity for people with dementia is detailed.

Collectively, these writings significantly contribute to our academic and clinical knowledge of assessing dementia. We have learned a great deal from studying and helping people with Down's syndrome; however, perhaps more importantly, this work should contribute significantly to our rather limited knowledge of assessing dementia in people with Down's syndrome and thus may step towards improving and widening access to service provision for these valued people.

CONTENTS

Acknowledgements 16

Author's declaration 17

Chapter 1 Introduction 18

1.1 Normal memory 20
1.2 Physical problems of ageing 28
1.3 Psychological problems of ageing 31
1.4 Problems with memory 36
1.5 Definition of dementia 38
1.6 Neuropsychology and clinical signs of Alzheimer's disease 41
1.7 Neuropsychology of Alzheimer's disease 50
1.7.1 Selective attention 51
1.7.2 Sustained attention 52
1.7.3 Working Memory Model 54
1.8 Neuropsychology of vascular dementia 59
1.9 Differential diagnosis 66
1.10 Learning disability and dementia 70
1.11 Neuropsychology of dementia with learning disability, and Down's syndrome 74
1.12 Social and cultural differences 80
1.13 Measuring dementia 84
1.14 Difficulties in assessing learning disability 91
1.15 Description and critique of instruments used 93
1.15.1 Wechsler Adult Intelligence Scale – Revised (WAIS-R) 93
1.15.2 Raven Coloured Progressive Matrices (RCPM) 99
1.15.3 Middlesex Elderly Assessment of Mental State (MEAMS) 104
1.15.4 Hampshire Social Services Assessment (HSSA) 108
1.15.5 Dementia Questionnaire for Mentally Retarded Persons (DMR) 112
1.15.6 Hospital Anxiety and Depression Scale (HADS) 117
1.15.7 Rationale and goals of this research 121
1.16 Outline of this thesis 124

PART I:	**Empirical Studies (Published Papers)**	172
Chapter 2	A neuropsychological test battery for identifying dementia in people with learning disabilities. S.B.N. Thompson (1994). British Journal of Developmental Disabilities, XL; 2 (79): 135-142.	173
Chapter 3	Examining dementia in Down's syndrome (DS): Decline in social abilities in DS compared with other learning disabilities. S.B.N. Thompson (1999). Clinical Gerontologist, 20; 3: 23-44.	194
Chapter 4	Rate of decline in social and cognitive abilities in dementing individuals with Down's syndrome and other learning disabilities. S.B.N. Thompson (2003). Clinical Gerontologist, 26; 3/4: 145-54.	241
Chapter 5	The Central Executive System in people with Down's syndrome and dementia. S.B.N. Thompson (2000). Clinical Gerontologist, 21; 3: 3-32.	261
PART II:	**Evaluation of Treatment and Services (Published Papers)**	322
Chapter 6	Design and evaluation of a computerised version of the Benton Visual Retention Test. S.B.N. Thompson, E. Ennis, T. Coffin & S. Farman (2007). Computers in Human Behavior, 23, 2383-2393.	323
Chapter 7	Improving visual memory with Aricept (Donepezil Hydrochloride, E2020) in mild-to-moderate Alzheimer's disease.	

S.B.N. Thompson, J. MacDonald &
T. Coates (2001).
Clinical Gerontologist, 24(1/2), 55-73. 336

Chapter 8 'Hollywood Wives': Education and support for spouses of inpatients with dementia.
S.B.N. Thompson & F. Watson (2001).
The Journal of Dementia Care, 9(2), 38. 357

Chapter 9 Interdisciplinary clinic for adults with early onset dementia in a mental health NHS trust.
S.B.N. Thompson, T. Coates, F. Chaâbane, P. Cherry, L. Collins, H. Pennicott, F. Watson & N. Rogan (2006).
Clinical Gerontologist, 29(4), 99-104. 361

PART III: Future Direction (Published Papers) 371

Chapter 10 Testamentary capacity and cognitive rehabilitation: Implications for head-injured and neurologically impaired individuals.
S.B.N. Thompson (2009).
The Journal of Cognitive Rehabilitation, 27 (Fall), 11-13. 372

PART IV: In Summary 382

Chapter 11 Discussion 383

11.1 Support and refutation of hypotheses 383
11.2 Suggestions for methodological improvements 397
11.3 Summary and conclusions of empirical studies 405
11.4 Future direction of research and key to improving services 409

Tables and Figures

Table 1.1: Differences between depression and dementia 33

Table 1.2: Similarities and differences between dementia and other physical and psychological problems 35
Figure 1.3: Adapted from Baddeley's (1992) Working Memory Model 56

Table 1.4: Intelligence classifications (adapted from Wechsler, 1981) 71

APPENDICES 425

Appendix A - A Note on the Mechanism of the Acetylcholine Esterase Inhibitors (ACIs) 426

Appendix B - Confirmation of Registration and Recommendation 429

Appendix C - Complete List of Publications in Topic Area 432

Appendix D - Papers Presented in Topic Area 438

Appendix E - Awards Gained in Topic Area 440

Appendix F - Summary of Experience in Topic Area 442

For
Moose and Bogey

ACKNOWLEDGEMENTS

I would like to thank all the clients and staff who participated in the research series that I conducted over the past years.

I am particularly grateful to the following for their helpful advice, support and faith in my pursuit of this Doctorate route: Marian Matthews (Academic Programmes Coordinator) and Dr Fiona Knight (Graduate School Manager) of the Graduate School, Bournemouth University; Professor Jim Roach (Dean) and Professor Mark Hadfield (Deputy Dean, Research & Enterprise) of the School of Design, Engineering & Computing (DEC), Bournemouth University.

My thanks also to Dr Sarah Hardy (Principal Clinical Psychologist) for diligently proof-reading my thesis and considering it to be worthy enough for external examination. Thanks also to Naomi Bailey (Research Administrator, DEC) for administrating the process so efficiently.

AUTHOR'S DECLARATION

This Thesis is submitted in partial fulfilment of the requirements for a Doctor of Philosophy by Publication degree at Bournemouth University.

I declare that this is my own work and does not contravene any academic offence as specified in the University's regulations.

Retention
I agree that, should the University wish to retain it for reference purposes, a copy of my Thesis may be held by Bournemouth University normally for a period of 3 academic years. I understand that my Thesis may be destroyed once the retention period has expired. I am also aware that the University does not guarantee to retain this Thesis for any length of time (if at all). I have been advised to retain a copy for my future reference.

Confidentiality
I confirm that this Thesis does not contain information of a commercial or confidential nature or include personal information other than that which would normally be in the public domain unless the relevant permissions have been obtained. In particular any information which identifies a particular individual's religious or political beliefs, information relating to their health, ethnicity, criminal history or personal life has been anonymised unless permission for its publication has been granted from the person to whom it relates.

Copyright
The copyright for this dissertation remains with me.

Requests for Information
I agree that this Thesis may be made available as the result of a request for information under the Freedom of Information Act.

Signed:

Name:

Date:

Chapter 1

INTRODUCTION

Increasing longevity, especially of people with a learning disability, has brought with it a seemingly ever-increasing demand on health and social services which in turn has seen an increase in research activity (eg Thompson, 2000). In particular, clinical psychology services in the United Kingdom (UK) have seen an increasing number of referrals for assessing older clients who have poor cognitive functioning, particularly Alzheimer's disease, and for providing advice for carers about clients who have declining memory ability (Thompson, 1993a). Supportive consultation with staff and clients alike is important and has increased the demands on all services as the size of the older population has grown.

In the UK, over 700,000 people have dementia (Milne, 2010). In the United States, Alzheimer's disease is the seventh leading cause of all deaths and increased 46.1 per cent between 2000 and 2006. In 2009, nearly 1 million families and other unpaid caregivers provided an estimated 12.5 billion hours of care to persons with Alzheimer's disease and other dementias, totalling $144 billion (Alzheimer's Association, 2010). More men than women have dementia, primarily because women live longer, on average, than men.

Identifying signs of declining memory and general cognitive functioning early on clearly has many advantages (Goldblum, Gomez & Dalla Barba, 1998), including the planning and provision of specialist care. Researchers and clinicians have been interested in the effects of ageing on the normal population for some considerable time (eg Holden, 1989) and have compared common impairments in short-term memory (Morris, 1996); psychophysiological differences and age-related memory decline (Young & Kramer, 1991). The difficulties of a differential diagnosis between depression and dementia have also been examined (de Groot, *et al.*, 2000a; O'Brien, *et al.*, 1996; Thompson, 1997; 2000) but the stumbling block of researchers has often been the transferability of measures to different client groups (Thompson, 2001a). Often standardised assessments are too difficult or are culturally-dependent; testing some clients results in floor or ceiling effects; and other tests are simply too demanding of a person's attention or concentration.

1.1 Normal memory

In order to understand the complexities of dementia, it is necessary to describe what happens in normal ageing and understanding what can go wrong and gives rise to abnormal conditions such as dementia. Ageing can be distinguished in terms of biological, social and psychological disciplines, but there is often a great overlap and interaction between them. For example, a physical change such as arthritis can limit mobility, which in turn can reduce involvement in social activities or other previous sources of enjoyment (Alcott, 1993). The influence of one aspect of ageing on another should also be remembered; this is important when considering and comparing past and present cognitive functions within the same person.

Defining 'normal' is a difficult task and it is surprising how 'normal' and 'abnormal' activities and attitudes often overlap. The blurring of boundaries occurs between different cultures, different environments or even between individuals. A misconception is to consider normality as distinct and opposite to abnormality when in fact 'normality' refers to the 'range around the middle of a dimension (eg height) with two extremes at opposite ends (very tall and very short), rather than one extreme' (Alcott, 1993). Different people have their own opinion about normality and hence expectancies in ageing are perceived differently between

individuals. With the advance of medicines and technology, people are generally living longer and so more people are exposed to older people and are witness to the variations in ageing of relatives and friends. In turn, people's understanding of normal ageing is being constantly revised and so too are their expectancies of themselves and others.

Normal ageing brings with it changes, not just to an individual's appearances, however subtle, but also certain changes to the higher mental functions or 'cognitive' functions (Allen, *et al.*, 1997; Freidl, *et al.*, 1997). Memory can also be affected (Craik, 1994; Small, *et al*, 1995), sometimes because the individual has failed to receive information correctly, or sometimes because it can no longer be effectively encoded or stored (Nyberg, *et al.*, 1996). The effect of ageing on memory, particularly episodic memory (Morris, 1994a), is very often one of the first of the cognitive functions to be noticed by others and can cause considerable distress to the individual and to relatives, close friends and carers. Deterioration in memory functioning is characteristic of dementia (Mitrushina, Uchiyama & Satz, 1995) but it can also indicate other dysfunctions which should always be considered in any assessment.

Memory functioning has been recognised as follows:

(i) Short-term and long-term memory – Short-term memory, now elaborated into the concept of working memory (Baddeley, 1992), is the system which allows one to remember a new telephone number while one is dialling it, so long as one is not distracted. Long-term memory allows one to remember a familiar telephone number from day to day and year to year (Collerton, 1993).

(i) Semantic and episodic memory – Different types of knowledge appear to be stored differently. A distinction drawn initially between episodic and semantic memories (Tulving, 1972) and, more latterly, a contrast between procedural and declarative memories has gained acceptance (Cohen & Squire, 1981). Episodic memories are for particular events, while semantic memories are context-free facts. For example, knowing what I had for breakfast is an episodic memory; knowing that the word 'breakfast' means a morning meal is a semantic memory (Collerton, 1993).

(ii) Declarative and procedural memory – Both semantic and episodic memories may be subsumed under this heading (which represent

the memory for facts). Procedural memory is for skills and routines and may include some types of sensory stimuli. For example, knowing how to drive a car is a procedural memory, knowing how the engine works is declarative (Collerton, 1993).

Generally, older people can learn as much as younger people (Fisk, *et al.*, 1997), but more time is needed for them to achieve the same level of learning as they cannot process and 'absorb' information as quickly as younger people (Salthouse & Meinz, 1995). Sometimes this speed reduction becomes noticeable and marked and may accompany the onset of depression (Krishnan, 1991). If memory has noticeably changed and continues to do so it can indicate the signs of a dementing process, if it is accompanied by another failure in cognitive function (APA, 1994).

Changes in language abilities can also be characteristic of dementia, but as an effect of the normal ageing process, people's voice characteristics tend to change with age with the pitch becoming higher at the fifth decade, the resonance thinner, and the volume lower (Alcott, 1993). Various factors, such as smoking, stooped posture, unclean environment (eg dust) or prolonged abuse of the voice can contribute to these changes. Ill-fitting dentures, toothlessness or weakening of the

muscles involved in speech production can all hinder speech and it is worthwhile investigating all practical aspects of a person's living environment and hygiene before drawing conclusions about a person's abilities or cognitive status.

Verbal skills, particularly the well-learned skills of reading, writing, vocabulary, and word usage, tend to be maintained (Moss & Patel, 1992); and the general intellectual status of healthy older people, as measured by neuropsychological tests, tends to remain within normal limits through the eighth decade (Simone & Baylis, 1997). Also, age-related decrement in semantic memory organisation and functioning are minimal if not absent (Goldblum, *et al.*, 1998). Arithmetic and memory tests that show decreased performance in older people, for example, Digits Backward of the Wechsler Adult Intelligence Scale - Revised (Wechsler, 1981), tend to reflect impaired concentration and mental tracking rather than decreased cognitive functioning (Parkin, Walter & Hunkin, 1995). Contrary to conventional belief, normal ageing processes do not affect the immediate memory span in older people (Nyberg, *et al.*, 1996).

Personality also plays a large part in normal ageing; some people adjust better than others to their change in circumstances, be it changes to their living environment, loss of occupational status, or physical changes

such as decreased mobility, lack of independent transport, and so on. Some individuals become more restless or agitated at the frustration of their changed world whilst others may be more placid or resigned and withdrawn (Thompson, 1997). Yet others adapt to change and are realistic about expectations and changes to their circumstances. Social adaptation and sexual changes are very often major causes of people's unhappiness. However, the general expectation that older people will not be sexually active (Schiavi & Rehman, 1995) is unfounded since there is a great deal of variation in both sexual interest and activity amongst all groups of people, young or older. Availability of a capable partner and acceptance of the level of a close relationship seem to be important factors in determining sexual activity or fondness. Exceptions are often around in most groupings and some older people never cease to amaze their younger relatives with the energy and wisdom sometimes not found in their younger peers!

Structural changes to the brain give rise to cognitive changes (Daigneault & Braun, 1993; Golomb, *et al.*, 1996) that may be seen by others observing the individual. In normal ageing, the brain undergoes several structural changes including decreasing in size, flattening of the surface, and increasing amounts of intracrannial space (Aylward, *et al.*,

1997; Deb, 1997; Kesslak, *et al.*, 1994; Prasher, *et al.*, 1996). Other microscopic and biochemical changes occur as well as changes of the EEG (Holland, 2000).

Normal intellectual decline associated with old age shows up most strikingly in four areas of intellectual activity; these can be summarized as follows:

(i) The primary, or working memory capacity of intact older people differs little from that of younger adults (Burack & Lachman, 1996) except when the amount of material to be remembered exceeds the normal primary storage capacity of six or seven items, as in tests of supraspan (Morris, 1996). Older people use less effective learning procedures - less elaborative encoding - and tend to show a greater differential between recall and recognition of learned material, particularly when the recognition tasks are easy (Fisk, *et al.*, 1997).

(ii) Diminished ability for abstract and complex conceptualization typifies the intellectual functioning of older people (Moss & Patel, 1992). The authors suggest that the more meaningful and concrete the presentation of a reasoning problem, the greater is the likelihood that people will succeed

at it.

(iii) Mental inflexibility, appearing as difficulty in adapting to new situations, solving novel problems, or changing mental set, characterizes intellectual performance failures of older age (Moss & Patel, 1995; Sattel, *et al.*, 1993). Some authors have suggested that apparent intellectual slowness in solving problems may be due to serial versus parallel processing. Evidence for slower serial processing was found in tests of older people as compared with younger participants (Ellis, Goldberg & Detweiler, 1996).

(iv) General behavioural slowing (Swearer & Kane, 1996) is a predominant characteristic of ageing that affects perceptual (Earles & Salthouse, 1995), cognitive (Sattel, *et al.*, 1993), and memory functions (Nyberg, *et a.*, 1996) as well as all psychomotor activity (Moss & Patel, 1992; Holland, 2000). Accurate evaluation of an older person's poor performance on any timed test must depend on careful observation and analysis of the effect of time limits on the scores, for the score alone will tell little about the effects of slowing per se (Lezak, 1983).

1.2 Physical problems of ageing

Confusion is commonly misunderstood to be a part of the dementing process. An acute confusional state is “a consequence of change in the body's metabolism which leads to high temperature, fever and delirium, which in turn can cause temporary disorientation, memory loss, a state of 'muddled perplexity', poor concentration, hallucinations, clouding of consciousness and restlessness”. (Goudie, 1993).

Unlike the situation where the person is suffering from dementia, the disorientation and confusion will improve if the underlying cause is treated. Regular check-ups are therefore important in ensuring that health problems and reactions to medication are dealt with before they lead to anything serious. Misdiagnosis can often occur in people who are over 65 years old mainly because certain reactions seem to indicate dementia at first glance. For example, acute confusional state can be caused by any of: poor diet, chest and urinary infections, heart disease, faecal impaction, sensory deprivation (eg poor eyesight, poor hearing, social isolation), grief reaction to bereavement, and so on. Signs such as changes in muscle tone, persistent language problems, perceptual problems (Earles &

Salthouse, 1995) and personality changes may indicate other conditions such as transient ischaemic attack (TIA) or a cerebrovascular accident ('stroke') (Thompson & Morgan, 1996). Haemorrhage in the blood vessels leading to the brain or in the vessels of the brain itself can result in a stroke. The cognitive changes associated with a stroke can be confused with a dementing process if the physical effects of the stroke are disguised or are subtle. Indeed, some small strokes do not cause devastating or obvious outward changes but many small strokes that cause death to specific brain sites (multi-infarcts) often lead to ‘vascular dementia’ (Krishnan, Hays & Blazer, 1997; O’Brien, *et al.*, 1998; Thompson & Morgan, 1996).

Lack of sleep can also effect a person’s cognitive functioning in that they are less able to concentrate and attend to tasks. Pollak and colleagues (1993) investigated sleep patterns in 29 elderly insomniacs. According to sleep logs, insomniacs took longer to fall asleep and stayed awake longer when they woke at night. A circadian rhythm of motor activity was found in both insomniacs and a control group with the former group being more active during periods of bedrest, which led to sleep deprivation and poor cognitive functioning. This has been explained in terms of shifts in the relative balances of aminergic and cholinergic modulation occurring

during waking and sleep (Sutton & Hobson, 1998).

These findings are interesting when considering Alzheimer disease patients who have disturbed cholinergic modulation (Kaufer, 1998; Qizilbash, *et al.*, 1998). Similar studies of circadian rhythm analyses conducted with Alzheimer disease patients have shown restlessness during sleep (Ghali, 1996) and an inability in some patients who have impaired endogenous pacemaker function to synchronize the rhythm of core-body temperature with the circadian cycle of rest-activity (Satlin, *et al.*, 1996). The control of circadian rhythms is thought to be a function of the suprachiasmatic nucleus of the hypothalamus (Cohen & Albers, 1991).

In a study of 16 patients diagnosed with dementia, Van Someren and colleagues (1997) showed that increasing illumination had stabilised the rest-activity rhythm for those patients with intact vision. In a later study of Alzheimer disease patients, Van Someren and workers (1998a), proposed that irregular day-night rhythm with behavioural restlessness during the night may be attributable to an underlying dysfunctional circadian timing system. Actigraphically obtained rest-activity rhythm of 14 different Alzheimer disease patients showed an improvement in its coupling of Zeitgeber after transcutaneous electrical nerve stimulation

(TENS) treatment but not after placebo treatment. In another study by Van Someren and colleagues (1998b), fitness training was shown to be helpful in elderly people suffering from sleep problems as circadian rhythm disturbances were again implicated. Swaab, *et al.* (1993) has hypothesised that improvements in behaviour of Alzheimer disease patients may be due to activation of the suprachiasmatic nucleus by light, mediated through the retina. Peripheral nerve stimulation in Alzheimer disease patients has been thoroughly investigated and discussed elsewhere (see Scherder, 1995).

1.3 Psychological problems of ageing

Depression is the most common emotional problem affecting older adults (Goudie, 1993). Even when the condition has been properly identified, many individuals do not receive treatment with antidepressants or are referred for specialist therapy, such as cognitive therapy (Blackburn & Davidson, 1990). Many older people adapt well to the times (eg changes in currency, government policies, and so on) and are able to reflect on the past in order to apply their experienced skills to the present day.

Identifying the signs of dementia and depression are key to treatment. Whilst it is generally not too difficult to list the signs of depression (APA, 1994, pp320-344, for example: low mood, loss of interest, sleep disturbance, weight loss, hopelessness, helplessness, thoughts of death or suicide, preoccupation with somatic complaints, agitation, loss of energy, feelings of worthlessness and guilt, thinking and concentration disturbances, forgetfulness), it is sometimes harder to distinguish between an older person suffering from depression alone, versus depression with dementia. Depression has been found to be associated with Alzheimer's disease in many studies (eg McDowell, 2001). However, some key points to sufferers of depression are also important in a diagnosis of dementia: forgetfulness; thinking and concentration disturbances; ability to maintain a task; lack of concentration. Thompson (1997) compares typical symptoms of depression with those of Alzheimer's-type dementia (Table 1.1).

Anxiety is also common and often overlooked in older people (Thompson, 1997). Typical symptoms include: 'butterflies' in the stomach, sweating, feelings of sickness, palpitations, and even diarrhoea.

TABLE 1.1: Differences between depression and dementia

(adapted from Thompson, 1993, p 8)

The person with depression	The person with dementia of the Alzheimer type
Often complains of a poor memory	Is often unaware of memory problems (later stages)
May say, "I do not know" in answer to questions which require thought or concentration	May 'confabulate' or make up answers to questions which require concentration or good memory and appears unaware that the answer is incorrect
Shows fluctuating ability and uneven impairment on cognitive testing	Tends to show consistent, global impairment on cognitive testing
Gives up easily, is poorly motivated and uninterested	Has a go
May be slow but successful in any complex task, aware of errors	Unsuccessful in carrying out tasks which require concentration, appears unaware of errors

Hyperventilation - breathing at a rate that is faster than normal - and dizziness, tightening of the chest, head and abdominal pains, can be the

result of an acute anxiety panic attack. Some sufferers of anxiety find that their arousal level is such that no one particular event or stimulus triggers their panic attack. This is termed 'free-floating anxiety' and can be difficult to treat but is claimed to be helped by practising relaxation regularly and exploring different ways of interpreting threatening or uncomfortable stimuli (Thompson, 1989). Anxiolytic drugs can also take the edge off severe anxiety and can help the sufferer explore new ways of coping.

There are of course several other conditions that might be confused with a diagnosis of dementia in older people. Some of these include paraphrenia, often psychosis related to alcohol problems (such as Korsakoff's), and Parkinson's disease (Goudie, 1993). Sometimes, the dementia-like symptoms are defined as 'schizophrenia of late life'. Thompson (1997) clearly illustrates the similarities and differences to dementia of several of the most common problems (Table 1.2).

TABLE 1.2: Similarities and differences between dementia and other physical and psychological problems (adapted from Thompson, 1997, p 12)

Problem	Similarities to dementia	Differences from dementia
Acute confusional state	Disorientation, poor concentration, self-neglect	Occurs rapidly, worse at night; disappears after underlying causes causes treated; clouding of consciousness
Depression	Poor concentration; slowness; non-responsiveness	Answers which but 'do not know' is frequent response
Anxiety	Inability to carry out day-to-day tasks because of agitation, catastrophic reaction - total failure to cope	No confabulation; insight into impaired functioning; when stressors minimized, ability is as normal
Paraphrenia	Misinterpretation of actions and statements, self-neglect	Some behaviour unimpaired, no missing out of steps in a task even if reasoning seems bizarre; hallucinations

1.4 Problems with memory

Memory failure is a common and significant problem in dementia (Greene, Hodges & Baddeley, 1995), hence it is important to first assess the extent to which it is a problem and for whom the problem is an obstacle.

It is now believed that there are four stages involved in memory: registration, encoding, storage and retrieval. For information to be stored in memory it must first be attended to or registered. Encoding is the process whereby this information may be semantically encoded or phonologically encoded (Baddeley, 1978; 1992), ie encoded in terms of meaning or sound, respectively. Storage is the process by which information is maintained in memory. It is widely accepted that different types of knowledge appear to be stored differently, so that, for example, knowing what a person ate for lunch (episodic memory) would be stored differently from knowing the word 'lunch' means a mid-day meal (semantic memory). Cohen and Squire (1981) have subsumed these terms under 'declarative memory' and reserve a further definition, termed 'procedural memory' for skills and routines including some types of sensory memory (eg knowing how to ride a motorbike is a procedural memory, knowing how the engine works is declarative). These functional definitions of memory have practical applications for therapists and are

also more simplistic than earlier definitions.

Retrieval is the process by which information is made available from memory and is thought to be dependent upon a number of factors, such as the closeness in which conditions are matched at encoding and retrieval, and the strategy used for retrieving memorized information (Thompson, 2001b).

It is important for clinicians and therapists to understand the mechanisms involved in memory functioning in order to be able to recognise and treat deficits when presenting as a consequence of dementia. Having an appreciation of the memory impairments has shown that people's intact memory is not always as comprehensive as one might imagine (Thompson, 1996); indeed, people's memory functioning can be very selective (Thompson, 1995). Selective memory is difficult to explain but generally people choose to remember only certain details of an event (Eich, 1984). People experiencing traumatic events sometimes repress distressing memories. The process used here is blocking the retrieval of information rather than preventing information from being memorized in the first place (Terr, 1994). Boring or over-complicated information is also selectively ignored and not retained. It is a constant battle, therefore, for therapists and clinicians to seek out stimulating information while still

achieving an objective assessment of a patient's memory functioning.

1.5 Definition of dementia

The definition of dementia generally accepted by clinical psychologists and psychiatrists is that outlined in DSM-IV (APA, 1994). In summary, it states that for a diagnosis of dementia, there should be demonstrable evidence of impairment in short-term and long-term memory. Impairment in short-term memory (ie inability to learn new information) may be indicated by an inability to remember three objects after five minutes. Long-term memory impairment (ie inability to remember information that was known in the past) may be indicated by an inability to remember past personal information (eg what happened yesterday; birthplace; occupation) or facts of common knowledge (eg past Prime Ministers; well known dates). The salient points of the full-length definition (all of which do not necessarily have to be present for a diagnosis of dementia) are:

1. Impairment of short-term and long-term memory;
2. Impairment of abstract thinking;

3. Impaired judgement;
4. Disturbances of higher cortical function (eg aphasia; apraxia; agnosia; constructional difficulty);
5. Personality change;
6. Specific organic factor;
7. Absence of a non-organic factor as a reason for the symptoms (eg major depression).

Dementia is commonly misunderstood to be a disease when in fact it is a syndrome, ie the result of a number of symptoms, and in a few instances it may be reversible. Stokes and Holden (1993) have described 'primary dementia' as an extensive, organic impairment of intellect, memory and personality. It occurs in the absence of clouding of consciousness (without drowsiness) which is acquired, irreversible and progressive.

Among people aged over 65 years old, the prevalence (ie the percentage of people afflicted at a given time) of moderate to severe dementia has been estimated at between 1.3 and 6.2 per cent (Stokes & Holden, 1993). The increased life expectancy of women, coupled with the greater prevalence of dementia in the ninth decade of life may mean that

more women than men suffer from Alzheimer's disease (Ruitenberg, Kalmijn & de Ridder (2001). However, after 90 years of age the incidence of Alzheimer's disease is higher for women than for men, but vascular dementia is higher for men than for women in all age groups (Ruitenberg, Ott & van Swieten 2001).

It has been common to distinguish 'presenile' dementia from 'senile' dementia both by age of onset and also by type of illness. Lishman (1987) in his text on Organic Psychiatry describes two types of dementia: arteriosclerotic (which may also occur as a presenile disease) and parenchymatous senile dementia. The latter, which refers to a dementing process in the 'parenchyma' or 'functional part' of the brain, is by far the commonest form of dementia and is generally characterized by those deficits found in Alzheimer's disease (Miller & Morris, 1993). Vascular dementia (Chui, *et al.*, 1992; Paul, *et al.*, 2001; Rhodin & Thomas, 2001; Román, *et al.*, 1993; Skoog, Kalaria & Breteler, 1999) is less common and, in the absence of Alzheimer's disease neuropathology, refers to the presence of small localized areas of dead tissue in the brain produced as a result of an inadequate blood supply.

Over the years, there have been several different definitions of 'dementia' and these have varied often according to the viewpoint of the

person proposing the definition; for example, from a neuroanatomist's structural viewpoint or from a neuropsychologist's functional viewpoint. Definitions have changed also with the advent of improved technologies such as Computerised Tomography (CT) scanning and Magnetic Resonance Imaging (MRI). Dementias resulting from a stroke, for example, are generally considered to be vascular dementia (Skoog, Kalaria & Breteler, 1999), or according to the structural defect, as in 'lacunar stroke'.

1.6 Neuropathology and clinical signs of Alzheimer's disease

Alzheimer's disease is named after a German physician, Alois Alzheimer, who first reported the disease in 1907. It is the single most common form of dementia, accounting for between 1% and 4% of the population per year, rising by half decade from its lowest level at ages 65 to 70 years to rates that approach 4% over the age of 85 years (DeKosky, 2001).

Initially, the neuropathology of Alzheimer's disease was thought to be arteriosclerotic; however, this was revised after researchers such as Corsellis and Evans (1965) and Tomlinson, Blessed and Roth (1970)

consistently reported no arteriosclerosis in people with a late onset Alzheimer's disease.

The clinical diagnosis of Alzheimer's disease is said to be correct 75% to 90% of the time (Morris, 1999). According to Dickson (2001), accuracy is highest for neurologists specialising in memory disorders and lowest for general practitioners, who have a tendency to overdiagnose Alzheimer's disease. The clinical accuracy also tends to be lower for older patients who often have mixed pathology rather than a single cause of dementia (Mendez, Mastin & Sung, 1992). The only clinical means of establishing a definite diagnosis is by microscopic examination of brain tissue as there are no laboratory tests and neither sophisticated imaging techniques nor detailed neuopsychological evaluation can specify Alzheimer's disease categorically (Dickson, 1999).

Typically, the onset is from 40 years of age onwards with insidious degeneration until death at about six years following onset (Lishman, 1987; Jorm, 1990; Burns & Levy, 1994). The brain invariably displays a degree of atrophy; however, age-associated atrophy and the normal variability in brain size preclude a diagnosis based solely on gross examination of the brain (Dickson, 2001). Atrophy of the medial portion of the temporal lobe is often disproportionate to other areas of the cortex.

In most cases, the primary sensory and motor cortices are relatively spared and on sectioning the brain, the lateral ventricles are usually dilated and the hippocampus and amygdala are atrophic (Dickson, 2001). More specific neuronal alterations accompany neuronal and synaptic loss. The most important of these alterations is paired helical filaments which are intraneuronal proteinaceous structures that are composed by an abnormal form of tau protein (see Cooper, *et al.*, 1995; Dickson, 1997; 1998; 2001; Dickson, Crystal & Bevona, 1995).

The neuropathological hallmarks of Alzheimer's disease is the intracellular neurofibrillary tangles of tau protein and amyloid plaques, primarily composed of aggregated amyloid beta (β) peptide. At high concentrations vesicular amyloid β aggregates to form high molecular weight species which are capable of seeding amyloid fibril growth. Hu and colleagues (2010) suggest that it is these aggregates that seed the extracellular amyloid plaque formation seen in the pathogenesis of Alzheimer's disease.

Studies of individuals in the general population with Alzheimer's disease, verified by neuropathology, have shown the clinical manifestations to follow three stages (Schneck, Reisberg & Ferris, 1982). The first involves a subjective opinion of forgetfulness which may be

accompanied by anxiety (Mohanaruban, Sastry & Finucane, 1989). The second is characterized by severe memory loss for recent events (Reisberg, 1983) with an impaired delayed recall being more pronounced than for immediate recall (Baddeley, *et al.*, 1991). Poor concentration, impaired orientation and minimal dysphasia are also usually evidenced (Rau, 1993), with vocabulary and memory for past events remaining largely unaffected. The final stage is marked by severe disorientation and cognitive abulia, ie absence or impairment of 'willpower' (Oliver, 1999; Oliver & Holland, 1986). Anxiety and other affective disorders are declining in the final stage (Reisberg, 1983).

There are also the effects of the residential setting on the elderly person (see Collacott, 1992). The level of staff support may vary according to the individual needs of the person. These factors become more important during planning decisions and in the provision of specialist care for individuals with a learning disability who are dementing.

Each person with Alzheimer's disease will vary slightly in presentation according to personality. Emotional, behavioural and cognitive changes will also vary, but generally accepted by clinicians and researchers is a stage model which describes broad characteristics

(Reisberg, 1983).

In the first phase, the 'forgetfulness phase', there is usually difficulty in recalling recent events, and a tendency to forget where objects have been placed (Greene, Hodges & Baddeley, 1995). Names of people and places, previously familiar, may be poorly recalled and a general disorientation persists and poor short-term memory (Goldblum, *et al.*, 1998). Abstract thinking, inability to concentrate on tasks and a marked lack of curiosity are also typical presentations and there may also be emotional changes such as anxiety and irritability and the 'new' or unexpected will be feared or disliked (Goudie, 1993; Thompson, 1997). Denial is also sometimes seen in presentation of people with Alzheimer's disease (Thompson, 1997).

Some researchers warn of the importance of accounting for attentional components in studies examining memory in Alzheimer’s disease; this is because memory and attention are interrelated cognitive processes that are most likely to influence the functioning of each other and yet they are difficult to distinguish in psychological experiments (Simone & Baylis, 1997).

The second recognised phase is known as the 'confusional phase'. Increasingly poor attention span and a decline in generalised intellectual

performance is seen with a deteriorating memory. Disorientation in place, word-finding difficulty and other changes to speech may be seen (Goldblum, *et al.*, 1998). Complex tasks are performed with difficulty, sometimes in a clumsy or inaccurate manner and often the skills the person learned last will be lost first. Hence the skills necessary for social independence and vocational skills are usually the first skills to be reduced or lost completely. Together with failing memory comes the concealment of these deficits by rationalising or confabulating events (ie providing an imaginary account of events or actions). Lack of interest in news and surroundings follows relatively quickly and can be extremely distressing to family and friends (Thompson, 1997).

The third phase, the 'dementia phase', is characterized by a lack of purpose in the person's behaviour which appears disjointed and sometimes bizarre. Remaining intellectual and self-care abilities require constant supervision as people in this phase undergo further deterioration in memory capacity, calculating ability (dyscalculia) and aspects of language are severely affected and eventually lost. Constant assistance is required for self-care skills such as grooming, dressing, toileting and for feeding. A progressive physical wasting can also be seen which will mean help with walking. Sometimes one or two years of life will follow in an

almost vegetative state until death.

Environmental factors may have a role in triggering Alzheimer's disease in susceptible individuals. An association between Alzheimer's disease and aluminium has been formulated for several years (eg McDowell, 2001). However, there is more compelling contradictory evidence (eg Flaten, 2001). There have also been studies in the past purporting the implication of cholinergic neurotransmitters, such as acetylcholine (Curran & Wattis, 1989). These studies were unconvincing until the late 1990s with the discovery of neurotransmitter pathways implicated in Alzheimer's disease and the subsequent trialling of the acetylcholine esterase inhibitors (ACIs).

These drugs are now the main treatment of choice in early onset Alzheimer's disease and include Aricept or *Donepezil Hydrochloride, E2020* (Kakinuma, *et al.,* 2010); Exelon or *Rivastigmine Tartrate* which is no longer prescribed widely because of lack of good efficacy results (Winblad, *et al.,* 2007; Kumar, *et al.,* 2008); Reminyl or *Galantamide Hydrobromide* (renamed Razadyne on 1st July 2005 because of the confusion with the diabetes drug Amaryl, manufactured by Sanofi-Aventis (Burns, *et al.,* 2008); and Ebixa or *Memantine* (Robinson & Keating, 2006) which acts on the glutaminergic (rather than the

cholinergic) receptor sites (see Appendix A – A Note on the Mechanism of the Acetylcholine Esterase Inhibitors (ACIs) on page 426).

Heredity appears to be of some importance to the risk of suffering from Alzheimer's disease. The identification of gene mutations and polymorphisms that either cause Alzheimer's disease or significantly increase the risk for developing it have enabled the creation of realistic rodent models of the disease (Chapman, Falinska & Knevett, 2001). However, whilst animals expressing mutated human amyloid precursor protein and presenilin-1 show dramatic parallels to Alzheimer's disease, as yet, none of the models appear to capture the full range of pathologies that characterise the human disease (Chapman, *et al.*, 2001).

The idea that genes can influence behavioural predispositions is becoming increasingly tractable (eg Isles & Wilkinson, 2000). In imprinted genes one allele is silenced according to its parental origin resulting in the inheritance of traits down the maternal or paternal line, in contrast to the more frequent mode of inheritance to the parental origin of the allele (Isles & Wilkinson, 2000). Hence, it has been suggested that genes may play an important determinant of behavioural outcome which might impact on such diseases as Alzheimer's disease.

As sporadic Alzheimer's disease is on the increase (Engelborghs & De Deyn, 2001) with growing demands on our medical services (DeKosky, 2001), there is a increasing need for early diagnosis. Cerebrospinal fluid (CSF) levels of protein tau have been shown to be significantly increased in patients with Alzheimer's disease (Andreasen, *et al.*, 1998; Andreasen, *et al.*, 1999; Hulstaert, Blennow & Schoonderwaldt, 1999). In recent years, it has become apparent that the β-amyloid component of senile plaques may be the key molecule in the pathology of Alzheimer's disease (Bayer, Wirths & Majtényl, 2001). Also, evidence has shown that the allele ε4 of apolipoprotein E (ApoE) is a genetic risk factor for Alzheimer's disease underlining the possible role of ApoE in the physiopathology of Alzheimer's disease (Hofman, *et al.*, 1997; Mahley & Rall, 2000; Merched, Blain & Visvikis, 1997).

By measuring CSF ApoE level, findings show that an increased level corresponds with an increase of mRNA ApoE in the brains of Alzheimer's disease patients (Merched, *et al.*, 1997; Nemes, *et al.*, 2001). Hence, Merched and colleagues (1997) concludes that CSF ApoE level seems to be a reflection of neuronal damage and/or an inflammatory reaction that may be common to Alzheimer's disease and other neurological and related diseases.

It would seem, therefore, that close relatives of a sufferer do have a greater risk of developing Alzheimer's disease (Myers & Goate, 2001). However, only a small per centage is due to gene mutations and apolipoprotein E4 is responsible for some 17 % of these cases (Fullerton, Clark & Weiss, 2000; Saunders, 2000; Wang, Kwon & Shah, 2000). The risk to relatives seems to vary depending on the age at which the disease began; and there is a decrease in risk with late age onset (Stokes & Holden, 1993).

Other risk factors, associated with Alzheimer’s disease, have also been reported, for example, diabetes mellitus (Luchsinger, Tang & Sung, 2001). However, further prognostic studies are needed (eg Ruitenberg, Ott & van Swieten (2001) and some would even advocate the use of routine screening of risk factors because of the increase in prevalence of Alzheimer’s disease (Milne, 2010).

1.7 Neuropsychology of Alzheimer's disease

In normal functioning, several types of information may be processed at once, possibly from a variety of sources. An example of this might be

holding a conversation or driving a car. These types of activities break down relatively rapidly in patients with dementia, even at the early stages. Alberoni, *et al.* (1992) reported that Alzheimer-type patients are particularly handicapped in keeping track of conversations involving several people. They also often have problems in remembering who said 'what' and 'when' (Thompson, 1997). Yet some studies have shown the preservation of certain skills such as the ability to recognise music (Wall, 2010) or even maintain past learned skills as a pianist (Beatty, *et al.*, 1999).

1.7.1 Selective attention

A specific component of executive processes (Baddeley, 1998), is selective attention (Morris, 1996). This has been most extensively investigated in relation to attentional shifts in spatial tasks (Simone & Baylis, 1997). Some researchers have distinguished between the three components that underlie attentional shifting, namely, engaging or focusing, shifting, and disengagement of attention (Morris, 1996).

In the auditory domain, deficits in selective attention may be

measured by the dichotic listening task (Mohr, *et al.*, 1990). Visual selective attention deficits may be evaluated by using visual display units to present stimuli via a screen (Filoteo, *et al.*, 1992). There is substantial evidence that shifting of attention is controlled, in part, by the posterior parietal region of the brain (LaBerge, 1990). Because of the substantial functional impairment in the posterior parietal cortex in Alzheimer's disease, a specific attentional deficit would be predicted and indeed, has been found to be the case (Parasuraman, *et al.*, 1992).

However, more recently, researchers have found that whilst patients with Alzheimer's disease have deterioration in both memory and attention, the progression of Alzheimer's disease is more closely related to deterioration of spatial memory (Simone & Baylis, 1997). The authors suggest that memory and attention are interrelated cognitive processes but are often difficult to distinguish in patient participant experiments.

1.7.2 Sustained attention

In the early stages of Alzheimer's disease, the patient is essentially 'alert' and able to take in information to a certain level of efficiency. At a

clinical level, 'alertness' readily translates into the cognitive concept of vigilance, the ability to detect a stimulus and respond readily (Morris, 1996). This is typically assessed using tasks in which a target occurs infrequently and unpredictably and has to be discriminated from other unpredictable items (Parasuraman & Giambra, 1991).

Interestingly, Alzheimer' disease patients were found to be no worse than controls on a tone discrimination task (Lines, *et al.*, 1991), but if the task complexity is increased, their vigilance becomes worse (Berardi, *et al.*, 1992). However, as a discrimination between non-Alzheimer's disease patients, such tasks do not appear to be of particular help clinically (Morris, 1996).

Clare, Whitaker and Nelis (2010) compared memory evaluations in healthy older people and people with Alzheimer's disease. Significant overestimation was found to be a frequent feature among people with dementia, with approximately two-thirds showing this pattern; although the authors also state that significant under-estimation is also reliably observed in a small proportion of people with dementia.

1.7.3 Working Memory Model

Information processing, attention and memory are successfully characterised by the Working Memory Model developed by Alan Baddeley and colleagues (Baddeley, 1986; 1992; 1996; Baddeley & Hitch, 1974). The model proposes a Central Executive System (CES) which functions to co-ordinate and schedule mental operations including the processing and immediate storage of information. This incorporates the notion of a 'scratch pad' system which holds and manipulates information simultaneously.

The CES has limited resources, hence the decline in performance associated with trying to combine two attentionally demanding mental operations. A cluster of peripheral systems support the functioning of the CES. This includes a specialised Articulatory Loop System (ALS), responsible for recycling verbally encodable information, such as when a person tries to keep in mind a telephone number for a short period or understand a grammatically complicated sentence.

In the visuospatial domain, there is the visuospatial scratchpad (VSSP) which maintains visuospatial imagery. These components of the model are interacting continuously with the CES devoting varying degrees of processing resources to each component depending on the task

(Fig 1.3). This is not the only model explaining sequencing and executive phenomena (eg Shallice, 1988; Furster, 1993), but it has provided a useful framework for studying cognitive impairment in dementia which has in part been characterised as a dysexecutive syndrome (Morris, 1986; Baddeley, *et al.*, 1986; Becker, 1988).

The peripheral systems of the Working Memory Model (Baddeley, 1992; 1998) are co-ordinated by the CES. The most extensively studied is the ALS which recycles verbal material in a relatively automatic fashion. For example, when remembering a string of digits, as in the digit span test of the Wechsler Adult Intelligence Scale Revised (Wechsler, 1981), the material is cycled continuously through the ALS to keep it in memory. A series of studies indicate that this system is unimpaired in Alzheimer's disease (Morris, 1984; 1987a,b; 1992). The evidence comes from experiments where patients are required to recall short lists of digits or words but the functioning of the ALS is suppressed by requiring them to mouth an irrelevant word (such as the word 'the'), effectively blocking articulatory rehearsal. 'Articulatory suppression' of this sort was found to have quantitatively the same effect on digits or words recall in Alzheimer's disease patients as in normal subjects. Thus, the equivalent

FIGURE 1.3: Adapted from Baddeley's (1992) Working Memory Model

THE CENTRAL EXECUTIVE SYSTEM

^	^
^	^
^	^
VISUOSPATIAL SKETCHPAD	ARTICULATORY LOOP SYSTEM
^	^
^	^
^	^

THE CENTRAL EXECUTIVE SYSTEM

loss of function without articulatory rehearsal shows the potency of the

system to recycle verbal material is undiminished in Alzheimer's disease (Morris, 1992).

The visual counterpart of the ALS is the visuospatial sketchpad which functions to retain visuospatial memory in immediate memory (Baddeley, 1992; 1998). For example, if a person's attention is diverted momentarily from a visual scene, they will still maintain the memory of that scene for a short period. This ability helps the person maintain a sense of visual continuity when, for example, moving around the room.

Although this ability has not been investigated extensively, there is evidence for significant impairment in visuospatial memory in Alzheimer's disease (Morris, 1994a). Firstly, patients with Alzheimer's disease have difficulty in a visual analogue of verbal span - the patient observes the experimenter tapping out a sequence on an array of nine blocks (or printed coloured squares as in the Wechsler Memory Scale Revised - Wechsler, 1988) in front of them and then has to immediately repeat the sequence of taps from memory. This block span performance has been found to be impaired consistently (Spinnler, *et al*, 1988).

Secondly, there is a measure known as the 'delayed matching to sample task' where an item is shown to the patient followed by a short delay followed by the same item with another or several others. The

patient is required to pick out the item seen before. By varying the delay it is possible to see how fast memory for the item decays. Sahakian, *et al.* (1988) used a version in which a pattern was shown on a computer screen and the patient had to identify the pattern from four others. Early Alzheimer's disease patients were found to show more rapid forgetting than normal older controls. A later study by Money, *et al*. (1992) found a similar impairment using filled circles of different sizes, but Alzheimer's disease patients were impaired when there was no delay between presentation and choice and showed the same rate of forgetting as the controls. Both studies, therefore, showed a substantial impairment in the performances of Alzheimer's disease patients.

Baddeley, *et al.* (1991) favours a localisationalist view; impairment in Alzheimer-type patients may be explained in terms of the dysexecutive syndrome and frontal lobe dysfunction. This links the notion of the CES to the Norman and Shallice (1986) model of attentional control which assumes that most ongoing actions are controlled by establishing routines. The routines or 'schemas' are mutually inhibitory and can be triggered by environmental events. Where they are insufficient to generate appropriate activity, a higher-level system, the Supervisory Attentional System (SAS) comes into play which is involved in coping with novel circumstances or

problem-solving activity (Morris, 1994b).

Shallice (1988) relates an impairment in the SAS to the difficulties patients with frontal lobe damage have in problem-solving. The conceptual link between the CES and the SAS has been introduced by Baddeley, *et al.* (1991), who suggests that an SAS impairment also results in the reduced capacity to direct and control attentional resources. Thus an impairment in the SAS may be closely analogous to the dysexecutive syndrome seen in Alzheimer's disease. Indeed, Alzheimer's disease patients do have damage to their frontal lobes (Morris, 1994b).

1.8 Neuropsychology of vascular dementia

Vascular dementia is a fluctuating and remitting vascular type dementia which is characterized by an abrupt onset (Stokes & Holden, 1993; Markesberry, 1998; Skoog, Kalaria & Breteler, 1999). The diagnosis demands neuropathology not showing Alzheimer’s disease changes and is defined in DSM-IV as follows:

‘A. The development of multiple cognitive deficits manifested by both

(1) memory impairment (impaired ability to learn new information or to recall previously learned information)

(2) one (or more) of the following cognitive disturbances:

(a) aphasia (language disturbance)

(b) apraxia (impaired ability to carry out motor activities despite intact motor function)

(c) agnosia (failure to recognize or identify objects despite intact sensory function)

(d) disturbance in executive functioning (ie planning, organizing, sequencing, abstracting).

B. The cognitive deficits in Criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.

C. Focal neurological signs and symptoms (eg exaggeration of deep tendon reflexes, extensor plantar response, pseudobulbar

palsy, gait abnormalities, weakness of an extremity) or laboratory evidence indicative of cerebrovascular disease (eg multiple infarctions involving cortex and underlying white matter) that are judged to be etiologically related to the disturbance.

D. The deficits do not occur exclusively during the course of a delirium.' (p 146).

Vascular dementia is the second most common form of dementia, after Alzheimer's disease (Skoo, Kalaria & Breteler, 1999). The aetiology, or cause of this type of dementia is a series of small strokes which may vary between individuals in frequency, intensity and also in location in the brain (de Groot, *et al.*, 2000a,b; Thompson & Morgan, 1996). Loss of specific cognitive functioning, (eg immediate memory functioning deficits, loss of visuospatial ability, attention and concentration deficits), minor neurological signs (such as weakness in the muscles on one side of the body, or slurring of speech) and sometimes periods of confusion may occur (Thompson, 1999a). Physical disability is usually not severe unlike that following a severe stroke. Following the infarct, there is usually

limited improvement until the next episode which can take place after a few weeks, months or even up to a year later (Thompson, 1999a).

The deterioration in cognitive functioning and mild disability is usually a step-wise process, as compared with Alzheimer's disease which is often an insidious gradual deterioration in functioning (Thompson, 1999a). However, recent studies have indicated that there may be a vascular connection with Alzheimer's disease (O'Brien, *et al.*, 1996; Rhodin & Thomas, 2001) in that cerebral vascular changes in patients with Alzheimer's disease probably precede the neuronal damage and dementia. Many people suffering from vascular dementia do not reach the end stage and die from a major stroke. However, early recognition and treatment of the underlying disease, such as hypertension, arteriosclerosis, or cardiac disease may inhibit further deterioration (Thompson, 1999a; Thompson & Morgan, 1996).

Researchers have attempted to devise methods for discriminating between Alzheimer's disease and vascular dementia such as CSF investigation (eg Nemes, *et al.*, 2001). However, determination of N^{ε}(γ-glutamyl) lysine concentration in CSF have so far not provided the discrimination available at post-mortem (Nemes, *et al.*, 2001). The clinical diagnosis of vascular dementia is similar to that of Alzheimer's

disease; indeed, they share the common risk factor, apolipoprotein E4 (Dickson, 2001).

In terms of cognitive functioning, patients diagnosed with subcortical ischaemic vascular dementia tested on recognition memory and verbal fluency performed better than Alzheimer's disease patients on the recognition memory tests (Tierney, Black & Szalai, 2001). Criteria for the diagnosis for ischaemic vascular dementia (IVD) has been made more rigorous by some groups of researchers (eg Chui, *et al.*, 1992; Román, *et al.*, 1993). Chui and colleagues (1992), for example, have drawn up a new set of criteria that describe 'probable IVD', 'possible IVD', 'definite IVD', or 'mixed dementia' (see Chui, *et al.*, 1992). Román and colleagues (1993), on the other hand, suggest that better interobserver agreement in the diagnosis of dementia was achieved from using the World Health Organisation International Classification of Diseases (ICD-10NA) (WHO, 1991). Diagnosis of dementia requires the presence of a 'decline in memory and intellectual abilities that causes impaired functioning in daily living' (Román, *et al.*, 1993).

'Impaired functioning in daily living' was accepted as a criterion for epidemiologic studies of vascular dementia because it would ensure that the changes are more than incidental and would increase specificity

(Román, *et al.*, 1993). Chui and colleagues (1992) also included 'interference with the conduct of the patient's customary affairs of life' as a requirement for the diagnosis of ischaemic vascular dementia. The impairment should be due to cognitive deficits and not to physical handicaps produced by stroke.

For the diagnosis of vascular dementia, Román and colleagues (1993) have suggested that cognitive decline should be demonstrated by loss of memory and deficits in at least two other domains, including orientation, attention, language-verbal skills, visuospatial abilities, calculations, executive functions, motor control, abstraction, and judgement. For diagnosis, memory deficits may not be as severe as in Alzheimer's disease, but single or isolated defects in cognition such as amnesic states, aphasias, and apraxias, do not meet the requirements. Therefore, although single cognitive deficits do not qualify,

> 'single lesions may produce vascular dementia when causing alteration of memory and at least two other cognitive functions of sufficient severity to cause impairment in daily living.' (Román, *et al.*, 1993, p 253).

Using the Mini-Mental State Examination (MMSE) (Folstein, Folstein & McHugh, 1975), Tatemichi, Desmond and Paitz (1991) have found 84% sensitivity, 76% specificity, a false-positive rate of 46%, and a false-negative rate of 6%, supporting the use of the MMSE for screening dementia stroke patients when adjustments are made to account for false positives. Other tests that could be useful for vascular dementia include the four-word memory test with 10-minute delayed recall, the cube-drawing test for copy, a verbal fluency test, Luria's alternating hand sequence or finger rings, the letter cancellation test for neglect, the reaction-time test, and the grooved pegboard test (Cummings, 1992; Grafman, 1991). The Mattis Dementia Rating Scale (Paul, *et al.*, 2001) has also been recognised as serving as a useful diagnostic tool in this respect.

To be considered as evidence in favour of vascular dementia, the radiological findings should fulfil minimum standards for both severity and topography (Román, *et al.*, 1993). In contrast, the California criteria (Chui, *et al.*, 1992), require 'two or more ischaemic strokes with at least one infarct outside the cerebellum'. Criteria from the National Institute of Neurological Disorders (NINDS) with support from the Association Internationale pour la Recherche et l'Enseignement en Neurosciences

(AIREN) (Román, *et al.*, 1993), recognise that a single lesion may cause vascular dementia and accept radiological lesions regardless of location as evidence of cerebrovascular disease. This picture is further complicated when considering a differential diagnosis (Dickson, 2001; Krishnan & McDonald, 1995; Rao, 1998).

1.9 Differential diagnosis

Sometimes the clinical picture resembles organic dementia yet there may be little or no indication of an organic cause. These types of disorders are termed 'pseudodementia' (Lishman, 1987). Often the distinction between organic dementia and pseudodementia is difficult to determine; as Lishman (1987) warns: '... in the early stages of organic brain disease a patient may occasionally react in such a way that his dementia is suspected of being more apparent than real - in other words a pseudodementia may turn out in fact to be a 'pseudo-pseudodementia'. ' (p 404). Only depression will be discussed here as it is the most salient factor in distinguishing between dementia.

In *depressive pseudodementia*, the patient most commonly becomes

slow to grasp essential aspects of the environment or about daily routines. Thinking is laboured and behaviour becomes inefficient because of difficulty in concentrating or because of inner preoccupations. Kemper and colleagues (1993) found that the greatest discriminant function between moderate Alzheimer's disease patients and those with pseudodementia was the simplest (versus more complex) of naming tasks, such as the WAB Responsive Speech task. Kemper and colleagues (1993) conclude that it is the Alzheimer's disease patient who does not know definitely how many days are in a week or other overlearned material.

The onset of endogenous depression is typically acute and recent, whereas that of dementia is insidious (Lishman, 1987). A careful history in depressive pseudodementia is therefore indicated and may well reveal that such capacities as memory have not been affected up to the time of presentation of 'dementia'. Patients with depression will often complain of their cognitive abilities in a way that is quite different to those with dementia. Usually these complaints are categorically stated and sometimes forcefully so. Lishman (1987) comments that there is often a tendency for depressed patients to counter questions by 'Don't know' responses rather than attempts to confabulate or make facile excuses for failure which is frequently observed in the patient who is organically

confused.

Several studies have warned of the difficulties of diagnosis especially in the early stages of dementia, as subtle personality changes are easily overlooked (Rossor, 1999). However, some distinctions can be made between the presenting clinical features of common conditions; for example, the onset of 'acute confusion' is usually sudden and of short duration; 'dementia' (chronic confusion) has a slow and steadily progressive onset, whilst depressive pseudodementia may take place over a number of discrete episodes (Goudie, 1993). Sufferers of depression seem generally to have insight into their condition, often complaining of an impaired memory; sufferers of dementia and acute confusion, on the other hand, seem not to have any insight into the problem (Mohanaruban, Sastry & Finucane, 1989). Other forms of pseudo-dementia include hypomania which can occasionally produce a picture which is mistaken for dementia. Pseudodementias are basically conditions in which a clinical picture resembles organic dementia yet physical disease proves to be little if at all responsible for the presenting symptoms (Mohanaruban, Sastry & Finucane, 1989).

Testimony of the difficulties encountered in the differential diagnosis of depressive pseudodementia are apparent (Benedict & Nacoste, 1990).

Reviews of follow-up diagnostic studies cite evidence of dementia being misdiagnosed as depression (Rossor, 1999). However, different opinions have been put forward concerning the nature of the depression observed in cases of Alzheimer-type dementia. Clinicians have conventionally viewed depression as a reactive phenomenon more likely to occur during the early stages of the dementia when a modicum of insight remains (Rossor, 1999). A similar explanation has been offered for the depression that accompanies vascular dementia (Rao, 1998; Román, 1999). DSM-IV (APA, 1994) recognises the frequent coexistence of depression.

Hurley and colleagues (2007) have attempted to identify the experiences of people with dementia who have poor or no communication. The authors developed a rating scale used by nursing staff to assess discomfort across nine quantifiable items. However, the pilot test has not been thoroughly investigated despite promising results indicating content validity. More reliable methods of assessing symptoms have included neuropsychological test batteries and the use of photon emission tomography (PET) scanning (Dubois, *et al.*, 2007; Nordberg, *et al.*, 2010).

1.10 Learning disability and dementia

Two per cent of the UK population (over one million people) have learning disabilities, the majority of them mild. In 1991, 4500 babies were born with severe learning disabilities (6 in 1000 live births); more people with learning disabilities are male (54 per cent) than female (46 per cent) (Mental Health Foundation, 1993).

'Learning disability' (or formerly, 'mental handicap') is a very broad term and has been used to describe people with an intelligence quotient (IQ) below 70. Wechsler (1981) has classified the abilities of groups of people according to IQ (Table 1.4).

People with learning disabilities commonly may have a range of difficulties which might include approaches to problem-solving, co-ordination difficulties, problems with speech or comprehension, cognitive delay, or slowness or inability to perform daily routines, such as hygiene or feeding (Thompson, 1993a). The range or number of difficulties an individual may have can be very large or equally, very small.

Increasingly, therefore, it has been useful to state a person's abilities rather than emphasising their negative disabilities. With the promotion of community living, definitions of learning disability have come to include

TABLE 1.4: Intelligence classifications (adapted from Wechsler, 1981)

Intelligence Quotient (IQ)	Classification
130 and above	Very Superior
120 - 129	Superior
110 - 119	High Average
90 - 109	Average
80 - 89	Low Average
70 - 79	Borderline
69 and below	Mentally Retarded

extent of a person's ability to live alone or his or her 'independence'. A

useful, working definition, taken from Thompson (1993b), has been adopted for several studies:

> ‘A person with a learning disability is someone who is, to a varying degree, dependent on others for their living needs because of a cognitive impairment resulting from hereditary abnormalities or directly following (or during) birth. They may (or may not) also have associated physical/sensory/behavioural/medical disabilities'. (p 195).

In DSM-IV (APA, 1994), “mental retardation” is defined as:

‘A. Significantly subaverage intellectual functioning: an IQ of approximately 70 or below on an individually administered IQ test (for infants, a clinical judgement of significantly subaverage intellectual functioning).

B. Concurrent deficits or impairments in present adaptive functioning (ie the person’s effectiveness in meeting the standards expected for

his or her age by his or her cultural group) in at least two of the following areas: communication, self-care, home living, social/interpersonal skills, use of community resources, self-direction, functional academic skills, wok, leisure, health, and safety.

C. The onset is before age 18 years.' (p 46).

'Down's syndrome' (previously termed 'mongolism') is the most frequently observed forms of learning disability (eg Moody & Moody, 1992). The former is used more often clinically and was first described by John Langdon Haydon Down as a separate entity in 1866 (Down, 1866) and independently in the same year by Seguin (Seguin, 1866). Seguin referred to the disorder as 'furfuraceous cretinism', emphasising an assumed relationship to cretinism, while Down, struck by some aspects of the physiognomy of the patients which were superficially similar to those of people in outer Mongolia, called it Mongolian idiocy. Thankfully, today people with such disorders are more commonly referred to by their first names, thus recognising the fact that there is a person behind such stigmatizing labels.

A characteristic of Down's syndrome is the presence of an extra chromosome ('trisomy 21') (Shermann, *et al.*, 1991). Often the person has developmental delays (Maclean, *et al.*, 1991), mostly a larger head circumference (Palmer, *et al.*, 1992), and language difficulties (Bigler, 1992; Pulsifer, 1996), especially with ageing (Young & Kramer, 1991). Down's syndrome has been the focus of much research and controversy (Barr, 1990); because of increased longevity (Eyman, Call & White, 1991), individuals with Down's syndrome are living long enough to be at risk for a host of age-related diseases (Evenhuis, 1997; Young & Kramer, 1991).

1.11 Neuropsychology of dementia with learning disability, and Down's syndrome

It should be noted that, just as with the general population, people with learning disabilities can develop any of the different types of dementias (Evenhuis, 1997). As with the wider 'normal' population, these different diseases have different courses, for example, step-wise versus insidious decline when comparing vascular dementia with Alzheimer's disease.

There is evidence to suggest that the neuropsychology of dementia in people who have Down's syndrome may be different (Pulsifer, 1996; Vincent, 1996), especially when considering the difference in level of performances obtained on neuropsychological tests (Cooper, *et al.*, 2001). However, these performances may depend upon the level of general abilities possessed by the client being tested.

It is interesting to note that it is an "overdose" of otherwise perfectly normal genes in Down's syndrome that causes disorders of human health, indistinguishable from major public health problems of the general population, such as Alzheimer's disease (Nizetic, 2001).

A number of difficulties arise when attempting to assess people with a learning disability, whether or not they possess the signs associated with dementia. For example, many of these clients have limited or poorly developed language (Bigler, 1992); poor comprehension; apraxia or agnosia; or suffer from depressive illness (Cooper & Collacott, 1993), or other psychiatric disabilities (Moss, Goldberg & Patel, 1991). They may have specific physical disabilities, such as incontinence, abnormal reflexes, or behavioural problems (eg inappropriate behaviour, stereotyped movements, or pronounced anxiety) that make conventional psychological testing awkward or even impossible. Without

electroencephalography (Devinsky, *et al.*, 1990) or brain scanning (Pinter, Eliez & Schmitt, 2001; Schapiro, Haxby & Grady, 1992), these clients may simply manifest the processes of normal ageing (Evenhuis, 1997).

Combining cognitive tests with other measures of dementia pathology, for example, brain scans (Pinter, *et al.*, 2001; Schapiro, Haxby & Grady, 1992), may reveal more distinctive early clinical indicators of cognitive deterioration. Although definitive diagnosis of Alzheimer's disease does appear to be forthcoming until post-mortem (Nemes, *et al.*, 2001).

In 1876, Fraser and Mitchell first noted an association between Down's syndrome and Alzheimer's disease, but it was not until 1929 when Struwe described the characteristic senile plaques of Alzheimer's disease in the brains of individuals with Down's syndrome (Cooper, *et al.*, 2001). Jervis (1948) and Verhaart and Jelgersma (1952) described clinical deterioration associated with Alzheimer-like changes at post mortem in a number of people with Down's syndrome; subsequent research focused on establishing similarities between the neuropathological changes in the brains of elderly Down's syndrome individuals and the senile plaques, neurofibrillary tangles and granulovacuolar degeneration characteristic of Alzheimer's disease. By the 1960s the link between the two disorders was

clearly established and now re-affirmed (Potter, 1991). However, the neuropathogical features and neuropsychology of people who have both Down's syndrome and Alzheimer's disease continues to be researched (Cooper, *et al.*, 2001; Evenhuis, 1997; Nizetic, 2001).

Currently, there are four genes that are implicated in risk for Alzheimer's disease (Pericak-Vance, *et al.*, 2000; Myers & Goate, 2001). Mutations in the genes that encode β-amyloid precursor protein (β-APP) (Goate, Charter-Harlin & Mullan, 1991), presenilin-1 (Sherrington, Rogaer & Liang, 1995), and presenilin-2 (Levy-Lahad, Wasco & Poorkaj, 1995) cause the rare early-onset form of familial Alzheimer's disease (see Grimaldi, Casadei & Ferri, 2000). The fourth gene, which encodes apolipoprotein E (Hofman, *et al.*, 1997), is a major risk factor in both early-onset (onset before 65 years) and late-onset (onset after 65 years) Alzheimer's disease (Corder, Saunders & Strittmatter, 1993; Scott, Grubber & Conneally, 2000). There are three common alleles of the ApoE gene: ε2, ε3 and ε4 (Myers & Goate, 2001). In Caucasian populations, individuals who carry the ε4 allele are three (heterozygotes) to eight (homozygotes) times more likely to develop Alzheimer's disease than individuals who do not harbour the ε4 allele (Corder, *et al.*, 1993).

However, there is still debate over whether or not people with Down's syndrome also present with typical clinical features of Alzheimer's disease (Oliver, 1999), since many such individuals maintain good physical and mental health into the fourth and fifth decades of life (Evenhuis, 1997).

Recent advances in genetics, however, allow us to better understand the link between Down’s syndrome and Alzheimer’s disease (Nizetic, 2001). Down’s syndrome, as a phenotypic result of trisomy 21, is a complex condition with a set of over 30 phenotypic features which manifest themselves with varying frequencies among affected individuals (Nizetic, 2001). The risk of Alzheimer’s disease among fathers of probands with Down’s syndrome has been found to be similar to that of control fathers (Schupf, *et al.*, 2001). Furthermore, Schupf and colleagues (2001) suggest that there is a fivefold risk of Alzheimer’s disease in mothers who gave birth to children with Down’s syndrome before age 35 which indicates a specific vulnerability to Alzheimer’s disease, as opposed to other age-related degenerative disorders.

There are difficulties in assessing people with Down’s syndrome because there may be other factors that contribute towards a poor performance on a neuropsychological test, for example, failure on a test

or poor performance might indicate dementia or an underlying disability (Atkinson, 1991a). Clearly, these restrictions would not be necessary if the 'intelligence' of this client population was evenly distributed throughout age groups (Oliver, 1999).

When considering these issues, other questions are raised. For instance, are the associated changes in behaviour present but undetected in people with Down's syndrome due to poor institutional environments? To complicate this further, it also seems likely that there is an increased prevalence of epilepsy with age in sufferers of Alzheimer's disease (eg Thompson, 1997). Making the distinction between the effects of long-term epilepsy and types of brain damage on cerebral function can often be a difficult process (Thompson & Morgan, 1996; Thompson, North & Pentland, 1992).

Although a high proportion of individuals with Down's syndrome develop the neuropsychological changes of Alzheimer's disease, only a proportion develop the definite signs of deterioration and have the clinical features characteristic of the later stages of Alzheimer's disease (Schupf, *et al.*, 2001; Thompson, 1997). It is also often difficult to discriminate between pre- or perinatal brain damage (eg meningitis; anoxia) in association with normal ageing and those considered to be the result of a

dementing process (Evenhuis, 1997). The situation is more complicated in people with a learning disability when there can be other confounding variables such as the long-term effects of institutional living, communication and comprehension difficulties, and the lack of a premorbid intelligence quotient since intellectual deficits may have originated from birth (Thompson, 1993b).

The paradox between unequivocal neuropathological findings and limited clinical evidence of dementia, particularly in Down's syndrome, has been partly resolved by the use of specific neuropsychological assessments to detect age-related deficits (Cooper, *et al.*, 2001; Pulsifer, 1996) but conclusive evidence to distinguish clinical features of dementia from normal ageing in people with Down's syndrome is still not available.

1.12 Social and cultural differences

In 1994, the 25th anniversary of the first statement of the principle of normalization in the human service literature was celebrated. Normalization, also termed 'social role valorization' (Wolfensberger & Kugel, 1969) is a complex term covering a number of important different

areas of living skills. Broadly, it can be defined in three ways:

(i) Values: Normalization is based on the belief that people with learning disabilities should be socially accepted and valued with the same rights as other non-learning disabled people who live in the mainstream of society as valued and respected citizens. Within different cultures, there are specific rights and societal positions. For example, the Chinese community is regarded as having a high respect for its elders, especially the grandparents, who have a key role in decisions and life in the family. Little is known about the integration of people with learning disabilities in specific cultures, but it is suspected that this varies greatly, with poorer care being associated with countries that have poor economies or poorly-run health services.

(ii) Health and social services: Normalization also has implications for the design and delivery of services to people with learning disabilities. There have been several changes in attitude over the years about the delivery and type of services, and management implications. For example, Local Based Hospital Units (LBHUs) used to be the chosen housing for people with learning disabilities moving from large institutions, into ward-type

accommodation often comprising 20- or 25-bedded dormitory-style rooms. A move away from LBHUs to smaller 'group homes' of 4 or 5 residents followed with an emphasis increasingly towards 'normal' accommodation such as houses or bungalows in ordinary housing areas. Older people, with or without learning disabilities, have often been re-housed according to financial constraints; some people moving from 'long-stay' institutions have been re-located to geographical areas previously unknown to themselves but with such tentative links as their original place of birth. Clearly, this is inhumane and people with learning disabilities deserve the rights given to everyone in choosing when and where to live.

(iii) Relationships: Normalization includes views and feelings about other people and how people with learning disabilities interact at a personal level. It is also about the rights of older people with learning disabilities and includes such rights as their sexuality (Thompson, 1994).

In 1985, Somerset Social Services Department and the local health authority put together an ambitious strategy (Turnbull, 1993); firstly, to establish social services as the lead agency in this field and secondly, to

close three hospitals for people with learning disabilities. The benefits for former residents, whose lifestyles were now more ordinary, and who were rightly proud of their achievements, was highlighted in an evaluation conducted by the University of Kent (Somerset County Council, 1992) that showed these new arrangements to be superior.

Around Great Britain, other agencies have evaluated their services following earlier recommendations from the Jay report (Jay, 1979) and Cullen (1991) report.

Services for people with learning disabilities in Gwent were provided for 10 years within the framework of the All Wales Strategy and All Wales Specialist Nurse Group (1992) and previously, the Welsh Office (1983). As Kay (1993) suggests, 'it took the Briggs Committee (Briggs, 1972) much prolonged discussion to suggest a new role for mental handicap nursing in 1972'. Indeed, it would seem that any change from existing services takes time and convincing the relevant decision-makers.

Certainly before long we will be faced with the situation of caring for older people with learning disabilities and with providing them with equivalent care to our existing older population. However, rather than training people with specialist roles, for example, specialist nurses, Cox (1993) and also Thompson (1997) advocate that we should be focusing on

meeting the various needs of people with learning disabilities; then, the role of the nurse will become evident. This seems to make sense so long as our specialism-trained staff are not lost or our special skills substituted by generic workers with rather diluted skills.

1.13 Measuring dementia

There are a number of assessment tasks available to the clinical psychologist; for example, the Rivermead Behavioural Memory Test (Wilson, Cockburn & Baddeley, 1991) is useful in establishing the level of a patient's procedural memory functioning but does not tell the clinician much about the patient's specific memory deficits particularly in which modalities the deficits may be occurring, ie visual or auditory. More specific cognitive testing using the Wechsler Memory Scale - Revised (Wechsler, 1988) allows for identification of visual or auditory memory deficits, the patient's ability to learn new items ("new learning") and visuospatial deficits.

If the patient has difficulty remembering particular words or people's faces, these deficits can be assessed using the Recognition Memory Test

(Warrington, 1984). Simple questioning about personal details, events and familiar routines is helpful to gain an impression of the patient's deficits and abilities. Interviewing spouse, close relatives, carers or friends about the patient's past history can be beneficial, and the Autobiographical Memory Interview (Greene, Hodges & Baddeley, 1995; Kopelman, Wilson & Baddeley, 1990) is useful for comparing information obtained.

Besides diagnostic uncertainty, there are a number of methodological issues that complicate the understanding of dementia, depression and their interaction. One of these is the issue that the course of many dementias is over a relatively long time span. Longitudinal studies are therefore favoured but these bring with them the problems of funding and resources, and natural attrition of subjects. There is also a problem of gaining consent for participation in research and permission from next of kin in the case of post mortem analysis (see Hagberg & Gustafson, 1985).

Applicability of test material that is already available and standardised, is also an important consideration. For example, the Test of Everyday Attention (Roberston, *et al.*, 1994) has been validated with Alzheimer's disease patients and those of low intellectual level. Transferability of norms across different client groups, for example, using the WAIS-R (Wechsler, 1981), has also been debated (Atkinson, 1991a).

Tests, such as the Middlesex Elderly Assessment of Mental State (Golding, 1989); Kendrick Cognitive Tests for the Elderly (Kendrick, 1985); Clifton Assessment Procedures for the Elderly (Pattie & Gilleard, 1979); or the Dementia Rating Scale (Mattis, 1988; Paul, *et al.*, 2001) are very useful screening tools for dementia and can indicate the need for further testing in specific areas of deficits, such as memory for faces, recognition of everyday objects or arithmetic ability.

Lishman (1987) warns that unless a full and comprehensive evaluation is made of a suspected dementing patient, the label of a primary dementing illness, for example, carries a hopeless prognosis. Care must be taken not only in carrying out tests but also in the interpretation of results. Hence, a specific and detailed knowledge base is required of a clinician to interpret results even if the actual tests have been carried out by non-specialised generic workers.

In recent years, there have been a number of studies seeking to measure the symptoms characteristic of dementia (eg Nagy, *et al.*, 1998; Schmand, *et al.*, 1998; Tyrell, *et al.*, 1996). Making a diagnosis of dementia, particularly in its early stages in a person with intellectual disability, can be a difficult process (Deb & Braganza, 1999). Following the evaluation of a screening instrument for dementia in ageing mentally

retarded people (Evenhuis, 1992; 1996), various studies have attempted to devise testing batteries for identifying dementia in individuals with intellectual disability (eg Burt & Aylward, 2000; Hon, *et al.*, 1999; Thompson, 1999b).

Some have suggested that the level and variability of intellectual disability have both militated against the use of existing neuropsychological tests and promoted the use of informant-based interviews (Oliver, 1999). Whilst others (Das, *et al*., 1995; Aylward & Burt, 1998; Mitchell, 1998) have identified and demonstrated the usefulness of individual assessments for people with dementia and learning disabilities.

A Working Group set up to devise such a testing battery recommended both administration of informant-based scales and direct assessment of the individual (Burt & Aylward, 2000). This was the conclusion of previous studies (Aylward, *et al.*, 1997; Burt, *et al.*, 1999). Further recommendations were that such a battery should include the facility for questioning informants who may be familiar with various aspects of the history and current functioning of the individual. This follows from the findings of Reiss (1987) and Gedye (1995). Such a scale for informing the diagnostic process is the Dementia Questionnaire for

Mentally Retarded Persons (Evenhuis, 1992; 1996) and the Hampshire Social Services Assessment (HSS, 1989) which rely on informant (carer) knowledge and completion. Information obtained from such scales add to the picture obtained from direct assessment of the patient (Deb & Braganza, 1999; Thompson, 1999b).

In terms of direct measures of dementia in people with learning disabilities, there have been very few reports in recent years. Das, *et al.* (1995) tested the Dementia Rating Scale (DRS) on moderately retarded subjects with and without Down's syndrome. Evaluation of the DRS as a measure of dementia was carried out by administering the following tests for a comparison of measures: the Peabody Picture Vocabulary Test (Revised), the Matrix Analogies Test (Expanded Form) and the Draw-A-Person Test. Results showed the DRS to be appropriate for clinical use as a screening tool for loss of competence due to ageing among individuals with mental retardation (Das, *et al.*, 1995). However, the subjects tested were moderately retarded; thus caution should be aired when testing more severely mentally retarded persons because of the possibilities of floor effects (Thompson, 1999b).

More recent studies (Hon, *et al.*, 1999) have examined the CAMCOG and its use as a neuropsychological assessment of older adults with

Down's syndrome. The advantages of the CAMCOG is that it yields separate scores on seven subscales as well as the total score. Hon, *et al.* (1999) tested a population of Down's syndrome individuals (age range 30 – 65 years) living in a single Health Authority catchment area. Of the 77 people with Down's syndrome in the area, 74 agreed to take part, making the study a near total population sample.

Scores on the CAMCOG were well distributed, with only 8 participants (11%) scoring zero on the test. This contrasted favourably with performance on the Mini Mental State Examination where there was a narrower range of scores and a higher percentage scoring zero. There was a significant difference in cognitive performance between younger (30 – 44 years) and older (45+ years) participants on the total CAMCOG score and on 6 out of 7 CAMCOG scales (Hon, *et al.*, 1999). The authors concluded that the CAMCOG, with minor modifications, is a useful test to assess those areas of cognitive function known to decline with dementia. Apart from those with pre-existing severe learning disability, severe sensory impairments and/or already advanced dementia, the authors stated that the participants were able to score above the floor of the test.

A similar direct assessment tool, devised by Golding (1989), is the

Middlesex Elderly Assessment of Mental State. This has the advantage over the CAMCOG of being simpler to understand and quicker to administer. Like the CAMCOG, it has separate scores for subscales as well as a total score, and does not appear to be subject to floor or ceiling effects (Golding, 1989; Thompson, 1999c). Although not proposed as a dementia screening tool *per se* without the accompaniment of other tests of cognitive function (Golding, 1989), it purports to measure symptoms of dementia over the same range as the CAMCOG. The Repeatable Battery for the Assessment of Neurological Status (RBANS) is a similar tool but is complicated to use because of its index structure and analysis (Schmitt, *et al.*, 2010).

Other validated tools have been used for assessing people with dementia with varying degrees of success. These have given rise to more discussion than consensus over their reliability, specificity and, of course, unsuitability of use with people who have a learning disability as well as dementia. Such measures include: the Geriatric Depression Scale (Hall & Davis, 2010); face-name associations (Hopper, *et al.*, 2010); California Verbal Learning Test (Lekeu, *et al.*, 2010); the Tower of London Test (Mrchegiani, Giannelli & Odetti, 2009); animal naming (Davis, *et al.*, 2010); and the Stroop Test (Balota, *et al.*, 2008). Tracking technology has

also been used by Oswald and colleagues (2010) for assessing outdoor mobility for mildly cognitively impaired people.

Environmental factors can influence assessment. Patients examined in a noisy setting, such as a room in a busy outpatient department or a part of the hospital that is particularly 'clinical' (has a clinical smell or plain walls), may perform less well (Thompson, 1995). Sometimes the patient is very disorientated and it may be necessary to establish the exact degree of disorientation the patient is experiencing; for example, the person may be unsure about the day or the month and year or does not know their date of birth or where they are being assessed. A checklist is often useful to determine this knowledge and can be used at frequent intervals to monitor the patient's level of orientation to their surroundings and circumstances. The Mini-Mental State Examination (Folstein, Folstein & McHugh, 1975) elaborates this type of questioning.

1.14 Difficulties in assessing learning disability

It should be noted that, just as with the general population, people with learning disabilities can develop any of the different types of dementias,

ie vascular, arteriosclerotic or parenchymatous (such as Alzheimer's disease). As with the wider 'normal' population, these different diseases have different course, for example, step-wise versus insidious decline when comparing multi-infarct dementia with Alzheimer's disease. There is no evidence to suggest that the neuropsychology of dementia is any different in the learning disability population excepting the difference in level of performances obtained on neuropsychological tests. These performances will depend upon the level of general abilities possessed by the client being tested.

A number of difficulties arise when attempting to assess people with a learning disability, whether or not they possess the signs associated with dementia. For example, many of these clients have limited or poorly developed language (Pulsifer, 1996); poor comprehension; apraxia or agnosia; or suffer from depressive illness (Cooper & Collacott, 1993), or other psychiatric disabilities (Evenhuis, 1997). They may have specific physical disabilities, such as incontinence, abnormal reflexes, or behavioural problems (eg inappropriate behaviour, stereotyped movements, or pronounced anxiety) that make conventional psychological testing awkward or even impossible. These clients may

simply manifest the processes of normal ageing (Kline, *et al.*, 2000; Zigman, *et al.*, 1996).

1.15 Description and critique of instruments used

When using any instrument of measurement for clinical evaluation, it is necessary to consider three key points: (i) reliability, (ii) validity, and (iii) standard error and norms. In addition, the use of some tests may have limited use with certain subjects because of (iv) floor and (v) ceiling effects. These points will be discussed with particular relevance to each of the tests administered.

1.15.1 Wechsler Adult Intelligence Scale - Revised (WAIS-R)

The Wechsler Adult Intelligence Scale - Revised (WAIS-R: Wechsler, 1981) provides the clinician with an overall 'intelligence quotient' or IQ, but more usefully, gives a profile of the patient's verbal and performance abilities on a variety of tests. The 11 subtests of the WAIS-R provide data

on a 'general intelligence' (g) factor, as well as two or three factorially derived group dimensions (Wechsler, 1981). Additionally, each subtest measures certain 'specific' or 'unique' abilities, capacities that are not assessed by the other subscales (Leckliter, Matarazzo & Silverstein, 1986).

Central to the diagnostic endeavour is a need to distinguish variation between subtests that can be attributed to measurement error and variation that reflects a true difference in underlying abilities (Atkinson, 1991a). McNemar (1957) published a table of reliabilities, standard deviations and standard errors of measurement of difference scores for the original WAIS (Wechsler, 1955). These statistics enable the clinician to determine the extent to which difference scores can be attributed to measurement error, and the extent to which they reflect true differences across ability levels. Piedmont, Sokolove and Fleming (1989) published figures for interpreting WAIS-R difference scores based on a sample of 229 psychiatric patients and recommended fuller exploration of the WAIS-R with more homogeneous clinical samples.

Atkinson (1991b) produced a similar table describing the properties of subtest score differences for the WAIS-R standardisation sample. He argued that calculations used by McNemar (1957) and Piedmont,

Sokolove and Fleming (1989) were technically incorrect for generalising beyond their samples since their standard error of measurement (which he claimed was purely descriptive in nature), provided an inflated estimate of score norms (Lord & Novick, 1968; Dudek, 1979). Atkinson's (1991a) data, based on a sample of 290 individuals with 'developmental delay', provides a better estimate; however, whilst potentially being a very useful tool, Atkinson (1991a) warns that even his sample is not presented as 'normative' for the learning disability population. He suggests that the tables provide a 'tentative empirical yardstick'. This still leaves clinicians and researchers with limited and non-generalisable norms with which to compare across people with a range of learning disabilities.

(i) Reliability

Since reliability is a function of the group on which it is determined, reliability information for the WAIS-R is provided separately by age group. Two type of data are offered: correlation coefficients and standard errors of measurement.

Different types of reliability computations were used to estimate the

reliabilities of the WAIS-R tests. When appropriate, a split-half procedure was employed (Wechsler, 1981). This produces a correlation coefficient between scores on two halves of the test, which is then corrected by the Spearman-Brown formula to obtain a reliability coefficient for the full length test. The split-half procedure is not appropriate for estimating the reliability of highly speeded tests (Digit Symbol) or tests where the two halves may be considered separate tests (Digit Span). For such tests an alternative method for estimating reliability is to use a test-retest procedure.

With the exception of coefficients for Digit Span and Digit Symbol, the reliability for each test has been determined using the split-half technique (usually odd versus even items) and correcting for the full length of the test. For Digit Span and Digit Symbol, reliability coefficients were derived from test-retest studies of samples at four age groups ranging in size from 48 to 80 individuals.

Across the age groups 35 years to 69 years, a high degree of reliability can be seen in the subtests; ranging from .67 as the lowest (Object Assembly: age range - 65-69 years) to .97 as the highest (Vocabulary: age range - 35-44 years; 55-64 years; 65-69 years). Of the overall IQ scores for the WAIS-R, the fullscale and verbal IQ is the most reliable, followed

closely by the performance IQ, across age groups. Respective correlation coefficients across age groups are: 35-44 years (.98; .97; .94); 45-54 years (.97; .97; .94); 55-64 years (.97; .97; .93); and 65-69 years (.98; .97; .94). Reliability coefficients of all WAIS-R tests can be seen in Table 10 of Wechsler (1981), page 30.

(ii) Validity

The initial evidence of the validity of the Wechsler adult scales stems from the procedures used to determine the content of the original 1939 Wechsler-Bellevue (W-B) Scale (Wechsler, 1981). Tests were selected for inclusion on the W-B based on their correlations with other established tests of intelligence and with empirical judgements of intelligence, on ratings by experienced clinicians, and on empirical studies of several groups of known intellectual level (Wechsler, 1958). Studies have been conducted relating Wechsler scale scores and various measures of academic success. In a concise summary of some of these studies, Zimmerman and Woo-Sam (1973) note that the strength of the relationship between Wechsler scale IQs and academic success is dependent on the adequacy of the latter. Comparisons of means for groups

of various levels of educational attainment consistently show that average Wechsler scale scores for individuals with lower levels of education are lower than scores for individuals with higher levels. Matarazzo (1972) draws the conclusion from a number of studies of various measures of intelligence, including the Wechsler scales, that the correlation between IQ and performance in school is about .50. Research has also shown the Wechsler adult scales to be related to various other correlates of global intelligence (Matarazzo, 1972).

(iii) Standard error and norms

In Wechsler (1981)'s manual, normative values are available for ages 16 to 74 years. ln Table 12 (p 33), he presents scaled scores on each of the eleven tests and the three IQs, fullscale, verbal and performance. To illustrate the interpretation of these statistics, at age 16-17 years the SE_M for Similarities is 1.29 scaled score points. This means that the chances are about two out of three that an individual's obtained score on Similarities lies within 1.29 scaled score points (one SE_M) of his or her "true" score, and the chances are about 19 out of 20 that the obtained score lies within 2.58 points (ie twice the SE_M) of the "true" score on

Similarities.

The standard errors of measurement reported by Wechsler (1981) are expressed in scaled score units for the eleven tests of the WAIS-R and in IQ units for the fullscale, verbal and performance IQs. This accounts for the fact that although the reliability coefficients are higher for the three IQs than for the separate tests, the standard errors of measurement of the IQs are uniformly larger than those of the eleven tests.

The average values of the SE_Ms presented for the eleven WAIS-R tests range from .61 scaled score points for Vocabulary to 1.54 scaled score points for Object Assembly. Information, Vocabulary and Block Design, which are the most reliable of the tests across all age groups, have the smallest standard errors of measurement; Object Assembly and Picture Arrangement, the least reliable of the tests, generally have the largest SE_Ms. For the three WAIS-R IQs, average SE_Ms are 2.53, 2.74 and 4.14 IQ points for fullscale, verbal and performance, respectively.

1.15.2 Raven Coloured Progressive Matrices (RCPM)

The 1956 edition of the Standard and Coloured Progressive Matrices and

the investigations reported in the Guide of 1965 (Raven, 1965), are the outcome of a group of studies begun at the Crichton Royal Hospital during the mid- to late- 1940s. The Raven Coloured Progressive Matrices (RCPM) are designed for use with young children and older people, for anthropological studies, and for clinical work. They can be 'used satisfactorily with people who, for any reason, cannot understand or speak the English language, with people who are intellectually sub-normal or have deteriorated' (Raven, Court & Raven, 1990; p 2).

The RCPM aim to measure logical thinking and problem-solving skills (Raven, 1965). The three sets of twelve problems constituting the coloured matrices are arranged to assess the chief cognitive processes of which children under 11 years of age are usually capable. The sets together provide three opportunities for a person to develop a consistent theme of thought, and the scale of thirty-six problems as a whole is designed to assess as accurately as possible, mental development up to intellectual maturity.

The coloured matrices are arranged to assess mental development up to the stage when a person is sufficiently able to reason by analogy to adopt the way of thinking as a consistent method of inference. This apparently decisive stage in intellectual maturation appears to be one of

the earliest to decline in later life, and the one most apt to be seriously impaired as the result of organic dysfunction (Raven, 1965; van den Broek & Bradshaw, 1994). Racial studies indicate that its muturation is partly a question of the native endowment of the individual, and partly the result of environmental influences and cultural opportunities, at least to the extent that in the absence of stimulation, the development of logical thinking tends to remain latent, or to develop somewhat later in life (Raven, 1965).

The way in which the test is presented, the fact that it is untimed, and the group of figures from which choice has to be made, have been chosen to ensure that success depends only upon a person's present capacity for intellectual activity. Presenting the test as coloured illustrations printed in a book makes the problem to be solved clear and with the least possible verbal explanation. Manipulation of the material is not essential for success, as a person need only indicate which figure they wish to insert in the problem to be completed. A person's maximum capacity for clear thinking has been found to vary with health and to improve with practice, less than their speed of accurate work (Raven, 1965). An untimed "capacity" test is therefore more useful than a test in which a person has to work against time. It has also been found that when the figures between

which choice has to be made are arranged below the problem to be solved, the distribution of choices is more uniform than is the case when the figures are arranged, for example, to the right (Raven, 1956).

Raven (1965) states that small groups of carefully selected subjects, rather than reliance of large numbers, were used in order to obtain diagnostically useful information. This meant that more information about individual subjects could be collected. However, the inevitable downfall of this policy is that comparison can only be made of relatively small numbers of people, for example, 50 healthy subjects.

(i) Reliability

A number of studies have assessed test-retest reliability of the RCPM across the world; for example, Li, *et al.* (1988) found a reliability of .95 used with normal Chinese children in Shanghai. Raven, Court and Raven (1995) report evidence from a study of 55 normal older adults in which there was a retest reliability of .79 following a six month interval. Other studies also support these latter findings (Measso, *et al.*, 1993; Vodegel-Matzen, van der Molen & Dudink, 1994).

(ii) Validity

Since its inception (see Raven, 1956), the RCPM has been further developed and various studies, particularly in the USA, have considered the validity of the RCPM (eg Emerling, 1990; Das & Jarman, 1991; Gainotti, *et al.*, 1992). These studies have led to the conclusion that three types of item can be identified within the RCPM: abstract reasoning by analogy, pattern completion through identity and closure, and simple pattern completion. Raven, Court and Raven (1995) report that in a German study of 180 subjects, these factors accounted for 36 per cent of the total variance. With a larger sample (783 children) in a Californian study, the same three factors accounted for 28 per cent of the total variance (Raven, Court & Raven, 1995).

(iii) Standard error and norms

Details about standard error for this test are not clear in the relevant literature, except for norms of children aged 5.5 - 11.5 years of age (Raven, Court & Raven, 1995; p 57), and for normal and emotionally disturbed children aged 6.5 - 12.5 years of age (Raven, 1965; p 39).

However, normative values of raw scores are stated, together with percentiles (Raven, Court & Raven, 1990; 1995: pp 37 - 43, and p 63, respectively). Additionally, qualitative comment can be found in Raven's (1965) guide about subjects who are "low-grade", "high-grade" and "seriously defective" intellectually. Respectively, these subjects find greater difficulty with the more complex figures and hence, generally score lower as the sets increase in complexity (set A1-12 is the easiest, then set A_b1-12, then set B1-12). Norms are only available for subjects aged 55 - 85 years (Raven, Court & Raven, 1995; p 63).

1.15.3 Middlesex Elderly Assessment of Mental State (MEAMS)

This test was developed as a screening test to detect gross impairment of specific cognitive skills in the elderly (Golding, 1989). It has been designed to assist clinicians to differentiate between functional illnesses and organically based cognitive impairments. Unlike a number of simple screening tests, for example, those intended to detect dementia, the MEAMS systematically surveys the major areas of cognitive performance using a comprehensive range of sub-tests, namely, orientation, name

learning, naming, comprehension, remembering pictures, arithmetic, spatial construction, fragmented letter perception, unusual views, usual views, verbal fluency, and motor perseveration.

The subject is required to perform a number of simple tasks, each designed to test some aspect of current cognitive functioning. The tests are sensitive to the functioning of different areas of the brain, each responsible for a different mental capacity (Golding, 1989). There are two versions of the MEAMS, version A and version B, to allow for reassessment. Each version comprises 12 subtests.

(i) Reliability

Various researchers have examined the reliability of the MEAMS (eg Golding, 1989; van Belle, *et al.*, 1990). In a study using 12 subjects with probable dementia and 12 matched controls, relative reliability of the MEAMS was found to be comparable to other tests used to assess cognitive function in dementia (Powell, Brooker & Papadopolous, 1993). Significant correlations ($p < .001$) were observed between trials 1 and 2 for the dementia group ($r = .82$) and across all subjects ($r = .95$). Relative reliability in the control group could not be assessed because on trial 1

nine control subjects achieved 100 per cent correct. Absolute reliability was examined by comparing the scores of each individual across trials. Subtest reliability was examined by discrepancies in the pattern of subtests passed or failed across trials. The discrepancies found between trials suggested a version effect; however, due to the small sample size this could not be verified conclusively. The authors concluded that the MEAMS was a useful and reliable global measure of cognitive function implying the use of the total score for comparisons rather than comparing individual subtest scores in the test-retest situation.

(ii) Validity

Results of past studies revealed that although the MEAMS was able to discriminate well between the organically impaired and other groups, it was not entirely satisfactory in its assessment of memory (Golding, 1989). For that reason, the MEAMS was supplemented with two additional subtests derived from the Rivermead Behavioural Memory Test (Wilson, Cockburn & Baddeley, 1985).

The principal validation of the MEAMS was based on older patients aged between 65 and 93 years of age who attended a London day hospital

(Golding, 1989). Four groups of patients were compared, each independently diagnosed with either multi-infarct or Alzheimer-type dementia (groups 1 & 2) and two sets of depressed patients (groups 3 & 4). Results indicated that performances by the dementia patients (groups 1 & 2) were substantially poorer on virtually every subtest than was the case for the patients diagnosed as suffering from depression (Golding, 1989). Furthermore, there was apparently no difference between the subgroup of patients diagnosed as depressive without the use of the MEAMS, and those who had subsequently been diagnosed in knowledge of their MEAMS score.

Results from validation studies have also shown a tendency for Alzheimer-type patients to be more impaired than multi-infarct dementia patients on orientation, naming, comprehension, spatial construction and perception of objects from usual and unusual views. It is difficult to know whether this pattern of results reflects a genuine difference in overall severity between the two conditions or simply some extraneous factor such as the point in the disease at which the patient is referred for assessment. Indeed, it is possible in Alzheimer disease for patients to show a more insidious onset than in multi-infarct dementia where the occurrence of a stroke may draw the patient to the attention of the general

practitioner.

(iii) Standard error and norms

Normative data (raw scores) across the subtests are available together with probabilities of occurrence. Total scores, means and standard deviations are also given from the data of 33 Alzheimer-type dementia patients, 25 elderly depressed patients and 60 elderly control subjects, ranging from 65 - 93 years of age. No norms are available for people with learning disabilities.

1.15.4 Hampshire Social Services Assessment (HSSA)

The HSSA is a carer-rated questionnaire to be completed about a patient whom the carer knows well or with whom the carer is in constant contact. The purpose of this assessment is to provide a general statement of the overall staff support level a person may need (HSS, 1989). Within this overall staff support level, there will be times and activities when support will be less or greater. High scores on the assessment reflect a higher level

of staff support required by an individual.

'Staff support' relates to providing training and other enabling opportunities in addition to the more traditional interpretation of 'support' as physical care of the patient. Weightings have been included in the assessment to incorporate support levels for particular skills teaching and physical care which are staff intensive. Scores attributed to individual items are not considered to be comparable to scores given to other individual items as weightings have been calculated for groups of items rather than individual ones.

Many of the items can also be found in other scales derived by Hampshire Social Services and Hampshire Health Authority, for example, the Hampshire Assessment for Living Independently (see HSS, 1989, for details). Scores are allocated according to level of independence and associated behaviours, communication and social skills. Weighting has been given to self-care skills, problem behaviours, social and communication skills, domestic/safety skills and leisure/community use skills.

The assessment yields a total score of 200 with 0 representing the lowest level of learning disability (ie the least support required) and 200 is the highest level (ie the patient requires the most support). Thirty-five

items in total are scored. Although several items include more than one skill area, weighting has been given approximately as follows to core skill areas: self-care (30%); social/communication skills (25%); domestic/home-based skills (25%); and leisure/community (20%).

The original sample population comprised 130 residents in Social Services accommodation for people with severe learning disabilities. This was drawn from a cross-section of the complete range of learning disabilities.

(i) Reliability

Twenty-six of the 130 residents approached in the original sample were assessed by a second assessor. When scores were compared, an inter-rater reliability coefficient of .84 was obtained which is significant at the 5% level. However, no test-retest reliability data is available.

(ii) Validity

Data were compared between assessors and those scores obtained from the officer-in-charge or keyworker responsible for the same patients. This

was carried out in order to compare the perceptions of each rater in assessing the patient's ability. A correlation coefficient of .71 was obtained ($p < .05$) for these comparisons. It is noted that this is not the same as considering whether or not a particular test measures what it is purported to do so, but it is instead a way of evaluating the success of the assessment in representing the "opinions" of raters about the "abilities" of their patients. The authors state: 'detailed tests of reliability and predictive validity are not relevant or appropriate for this assessment. Scores give snapshots of independence ratings in one particular environment' (HSS, 1989; p 8).

Although this statement is debatable on the grounds that any instrument should be shown to be both reliable and valid, acknowledgement is made that there may be many factors involved in arriving at a score for a particular patient in a particular setting and that it is sometimes difficult or impossible to replicate conditions and so demonstrate these points. There is usually a way of demonstrating that a measure shows the same thing given the same setting conditions; this is sometimes achieved by comparing it with similar measures. However, there does not appear to be anything similar with which to compare it with to date.

(iii) Standard error and norms

No raw data or standard errors are available. Scores (0 to 200) are shown in 6 bands: 0 - 30; 31 - 60; 61 - 110; 111 - 150; 152 - 200; 200+ (HSS, 1989; p 7). Age range of sample was 30 - 70 years.

1.15.5 Dementia Questionnaire for Mentally Retarded Persons (DMR)

To facilitate the diagnosis of dementia in 'intellectually disabled persons', the DMR was devised by Evenhuis (1992a). This is an English translation of the former identical Dutch questionnaire published in Evenhuis, Kengen and Eurlings (1990). The DMR is a standardised informant-based questionnaire consisting of 50 items to be completed by family or staff about the patient who is known to them. Evenhuis (1992a) suggests that the items are comparable to questions put by a physician, psychologist or psychiatrist to a patient or to a carer about a patient. They are based on knowledge of symptoms of dementia obtained from the American Psychiatric Association (APA, 1987), and on the authors' experience with

the behaviour of intellectually disabled people. Although items are placed in the questionnaire in an arbitrary sequence, to prevent response tendencies, they can be arranged into 8 subscales (divided into two categories): (i) cognitive scores: short-term memory; long-term memory; spatial and temporal orientation; and, (ii) social scores: speech; practical skills; mood; activity and interest; behaviour and disturbance.

The DMR is provided with a simple linear scoring system. The items have three response categories: 0 points = no deficit, 1 point = moderate deficit, 2 points = severe deficit. Therefore, higher scores correspond to more severe deterioration. The subject's behaviour during the last two months has to be judged. If an item cannot be judged, for example, in the case of a lack of expressive capacities of the subject, then the score has to be "2". The questionnaire is provided with a short instruction and takes about 15 - 20 minutes to complete.

The DMR has been evaluated in a number of studies (Evenhuis, Eurlings & Kengen, 1984; Kengen, *et al.*, 1987; Evenhuis, 1990; Evenhuis, 1992b). Inter-rater reliability, internal consistency of items, relationship between intellectual level and scores, influence of some physical handicaps on the scores, relationship between the diagnosis of dementia and scores, and the relationship between the diagnosis of

depression and scores was investigated in two cross-sectional studies among older residents of three Dutch institutes (Evenhuis, 1990; 1992b). Additionally, in the first of these two prospective longitudinal studies (Evenhuis, 1990), during 1985 - 88 in 17 middle-aged institutionalised subjects with Down's syndrome, the relationship between the expert's diagnosis of dementia and score changes was investigated.

In the second study during 1983 - 89 in 139 older institutionalised subjects without Down's syndrome (Evenhuis, 1992b), again, the relationship between the expert's diagnosis of dementia and score changes was examined, to develop provisional criteria for interpretation of DMR score changes. In this study, absolute scores were also analysed and provisional criteria were formulated for a diagnosis based on a single completion of the DMR. In a final longitudinal study during 1988, again reported in Evenhuis, 1992b, the predictive value of the provisional criteria was tested both in residents aged 70+ years without Down's syndrome and in residents aged 35+ years with Down's syndrome.

(i) Reliability

Results of examining inter-rater reliability (Evenhuis, Eurlings & Kengen,

1984) revealed correlations for the following items: short-term memory (.84); long-term memory (.87); orientation (.86); speech (.68); practical skills (.94); mood/activity and interest (.74); behaviour disturbance (.44). The last correlation (.44) apparently was caused by one of the six pairs of raters. No test-retest reliability data was obtained.

(ii) Validity

Evenhuis, Eurlings and Kengen (1984) state that items that 'correlated insufficiently with the other items within their subscale, were removed'. T-tests that showed those with a diagnosis of dementia scored significantly higher than the group with a diagnosis of 'no dementia' on the subscales short-term memory, orientation, speech, practical skills and mood. This finding was confirmed by discriminant analysis. When scores of individuals were classified according to the results of discriminant analysis, in 72% of subjects a DMR diagnosis was made, corresponding to the diagnosis. A correct diagnosis based on DMR scores seemed particularly difficult cases of a low intellectual level (severely or profound), extreme apathy, or clouded consciousness. With regard to the diagnosis of depression versus dementia, results from two studies

(Evenhuis, Eurlings & Kengen, 1984; Kengen , *et al.*, 1987), found no relationship between a diagnosis of depression and scores on the subscales "mood" and "activity and interest".

(iii) Standard error and norms

Standard errors are not mentioned in the Evenhuis' (1992a) manual but a table of norms is presented enabling a comparison to be made between 'level of retardation' (IQ score), age, total scores, and changes in total scores following test-retest. In summary, score changes in the total of scores of items comprising the cognitive subscale (ie 'sum of cognitive scores' or 'SCS') give a more favourable sensitivity-specificity relationship than do score changes of the social subscale (ie 'sum of social scores' or 'SOS') (Evenhuis, 1992b). Independent of the level of 'retardation', an increase of 7 points in the SCS and/or an increase of 4 points in the SOS has been found to be indicative of dementia (Evenhuis, 1992a), using the American Psychiatric Association's (APA, 1987) criteria for the diagnosis of dementia.

1.15.6 Hospital Anxiety and Depression Scale (HADS)

The HADS is a self-assessment scale for detecting states of depression and anxiety in hospital outpatients. First presented by Zigmond and Snaith (1983), the scale comprises 14 questions offering a 4-choice response to each question ranked 0 - 3. For example, question 1 asks if the respondent feels tense or 'wound up' and offers the choice of responses: 'Most of the time', 'A lot of the time', 'From time to time, occasionally', and 'Not at all'. The ranks can be totalled to give a score for 'anxiety' related questions and a total score for 'depression' related questions. A total score in either category within the range 0 - 7 suggests that the respondent's score is in the 'normal range'; a score falling in the range 8 - 10 is 'borderline', and in the range 11 - 21 is 'clinical' or "caseness" for anxiety or depression.

Zigmond and Snaith (1983) have tested the use of the HADS scale in outpatients aged 16 to 65 years of age attending general medical clinics. The authors state that in their opinion 'there is no reason to suppose that its use would be invalid in patients attending other hospital clinics but further research is needed ...' (p.366). Results from the use of other scales used for the diagnosis of psychiatric conditions were compared, for

example, the General Health Questionnaire (Goldberg, 1972; Goldberg & Hillier, 1979), and various depression rating scales (Kearns , *et al.*, 1982).

(i) Reliability

Following the accumulation of data on 100 subjects, the reliability of the HADS was tested (Zigmond & Snaith, 1983). There were found to be 1% false positives and 1% false negatives (depression subscale), and 5% false positives and 1% false negatives (anxiety subscales). These figures indicate the number of occasions a type I or type II error have occurred (eg if a diagnosis of depression is made in the absence of depression; or if a diagnosis of 'no depression' is made in the presence of depression; etc). However, test-retest data is not known.

The internal consistency of the two subscales (anxiety and depression) was examined by calculating correlations (Spearman) between each item and the total score of the remaining items in the subscale (Zigmond & Snaith, 1983). For the anxiety items, the correlations ranged from +.76 to +.41 and the significance of all these was $p < .01$. The analysis of the depression scale items revealed one weak item ($r = .11$), which was removed, and a range of correlations from +.60 to +.30 ($p < .02$). Hence,

overall there was a high degree of internal consistency amongst items. Further research (Zigmond & Snaith, 1983) also revealed that the subscales were both reliable and valid in being used as measures of severity.

(ii) Validity

Zigmond and Snaith (1983) examined whether the anxiety and depression subscales detected different aspects of mood disorder or, alternatively, were so closely related that they could be considered to be estimating much the same thing, for example, a general index of emotional disturbance. In checking this, the authors noted that, in clinical practice, many patients suffer from similar degrees of both anxiety and depression, and that this was indeed the case with the sample tested. It was therefore clear that high correlations between estimates of anxiety and depression would be expected in any sample containing a high proportion of patients with similar degrees of both disorders.

In order to overcome this, the authors selected from their sample all those patients in whom there was a distinct difference between the interviewers' assessments of the severity of anxiety and depression. They

examined the data of those patients in which there was a difference of two or more points in the severity; there were 17 such patients. The results of this analysis revealed that, whereas the patient-rated subscales correlated significantly with the interviewers' assessments of the appropriate mood disorder (anxiety: +.54; depression: +.79), there were significant correlations between the contrary disorders. Although the size of the sub-sample was small, these findings give some support to the view that the subscales do in fact assess different aspects of mood disorder.

A further investigation considered whether or not the subscale scores were influenced by physical illness, apart from mood disorder. Using subjects matched for age and sex with the normal sample, the differences were tested using the Students t-test. For both subscales, the scores yielded non-significant results. Hence, the physically ill patients of their study, (who were assessed as not having a mood disorder), had similar scores to the normal sample; this enabled the authors to conclude that the scale scores had not been affected by the respondent having a physical illness.

(iii) Standard error and norms

No standard errors are quoted in Zigmond and Snaith (1983). The normative ranges for scores on the HADS for ages 16 to 65 years are: 0 – 7 ‘Normal’; 8 – 10 ‘Bordeline’; and 11 – 21 ‘Clinical’.

1.15.7 Rationale and goals of this research

There is substantial evidence to show that cognitive abilities may decline with the onset and progress of dementia. A link has been found between people who have the neuropathology of Down's syndrome and their subsequent development of clinical signs, associated with Alzheimer's-type dementia. Less is known about the effects of dementia on non-Down's syndrome persons with learning disabilities and whether there is any difference when compared with Down's syndrome individuals.

Comparing cohorts from other studies that have examined dementia in people with learning disabilities (eg Crayton & Oliver, 1993), a five- to six-month period between test and retest is quite typical and generally acceptable. In fact, a twelve-month follow up was chosen in order to

maximize any possible changes and effects, such as cognitive decline.

Goal 1

To compile a neuropsychological test battery to identify dementia in people with Down's syndrome.

Goal 2

To compare the social decline in dementing individuals who have Down's syndrome with other groups of people with learning disabilities.

Goal 3

To compare the cognitive decline in dementing individuals who have Down's syndrome with other groups of people with learning disabilities.

Goal 4

To examine the Central Executive System theory in the context of the

neuropsychological research findings.

Goal 5

To explore the use of a computerised version of the Benton Visual Retention Test for testing memory functioning.

Goal 6

To examine the potential neuropsychological benefits of prescribed medication (Aricept) in non-Down's syndrome adults.

Goal 7

To evaluate the use of a support group for carers of spouses with dementia.

Goal 8

To evaluate the use of an interdisciplinary clinic for assessing adults with

early onset dementia.

Goal 9

To explore the legal standpoint on testamentary capacity and produce recommendations for improvement.

1.16 Outline of this thesis

Theoretical considerations about the difficulties of assessing people with learning disabilities, particularly people who have Down's syndrome and dementia, continue to be discussed throughout this thesis. Importantly, these issues concern intellectual development and impact on the cognitive rehabilitation and integration into the community of such individuals.

Part I: Empirical Studies (Published Papers), chapters 2 – 5 explore the empirical findings from clinical studies and address research Goals 1-4. All clients participating in the studies were randomly selected from consultant psychiatrists' lists of people living in their own home or in voluntary sector group homes in England.

In Chapter 2, a neuropsychological test battery, devised to identify dementia in people with Down's syndrome, is discussed. In Chapter 3, decline in social abilities, often associated with dementing adults, is measured and compared between those with Down's syndrome and those who have other forms of learning disabilities. In Chapter 4, the rate of decline in cognitive abilities is also considered together with social abilities in people with Down's syndrome or another form of learning disability. In Chapter 5, the Central Executive System theory is examined in the context of neuropsychological findings from testing individuals who have Down's syndrome and dementia.

Part II: Evaluation of Treatment and Services (Published Papers), chapters 6-9 consider ways of improving services designed to assess people with dementia and address research Goals 5-8. Chapter 6 explores the use of a computerised version of a well-used and standardised test of memory functioning. In Chapter 7, the most frequently prescribed "anti-dementia" medication, Aricept, is examined for its potential benefits for memory functioning in mild-to-moderate Alzheimer's disease. In Chapters 8 and 9, new services are examined that provide assessment and support for people with cognitive difficulties arising from dementia.

Part III: Future Direction (Published Papers), chapter 10 considers the

legal aspect of Wills and provides suggestions for improving the legal test for testamentary capacity.

Chapter 11 brings together the empirical findings in a discussion and conclusions. Treatment efficacy and the provision of services is discussed and lessons learned from the research studies.

Conducting the clinical studies has enabled significant suggestions to be made for future research and support services. As well as consolidating these results and integrating them into our existing knowledge of dementia, important conclusions are made that contribute and extend our present knowledge of both Down's syndrome and the complex nature of clinically assessing dementia in the general population.

REFERENCES

Alberoni, M., Baddeley, A.D., Della Sala, S. & Logie, R. (1992), 'Keeping track of a conversation: impairments in Alzheimer's disease', *International Journal of Geriatric Psychiatry, 7:* 639-646.

Alcott, D. (1993), 'Normal Aging' in Stokes, G. & Goudie, F. (eds),

Working with Dementia, Oxford: Winslow Press, 9-15.

Allen, P.A., Smith, A.F., Jerge, K.A. & Vires-Collins, H. (1997), 'Age differences in mental multiplication: Evidence for peripheral but not central decrements', *Journal of Gerontology,* 52; 2: P81-P90.

All Wales Specialist Nurse Group (1992), *'A Statement for the Future: Mental Handicap Nursing in Wales',* Cardiff: Welsh Office.

Alzheimer's Association (2010), 'Alzheimer's disease facts and figures', *Alzheimer's & Dementia, 6;* 2: 158-194.

Andreasen, N., Minthon, L., Clarberg, A., *et al.* (1999), 'Sensitivity, specificity, and stability of CSF-tau in AD in a community-based patient sample', *Neurology, 53:* 1488-1494.

Andreasen, N., Vanmechelen, E., Van de Voorde, A., *et al.* (1998), 'Cerebrospinal fluid tau protein as a biochemical marker for Alzheimer's disease: A community based follow up study', *Journal of Neurology, Neurosurgery & Psychiatry, 64:* 298-305.

APA - American Psychiatric Association (1987), *'Diagnostic and Statistical Manual of Mental Disorders - Revised (DSM-III-R). 3rd ed',* Washington DC: American Psychiatric Association, 78-80.

APA - American Psychiatric Association (1994*), 'Diagnostic and Statistical Manual of Mental Disorders (DSM-IV). 4th ed',*

Washington DC: American Psychiatric Association, 139-143.

Atkinson, L. (1991a), 'Mental retardation and WAIS-R difference scores', *Journal of Mental Deficiency Research, 35;* 537-542.

Atkinson, L. (1991b), 'On WAIS-R differences scores in the standardization sample', *Psychological Assessment: A Journal of Consulting & Clinical Psychology, 3;* 288-291.

Atkinson, R.C. & Shiffrin, R.M. (1968), 'Human Memory: A Proposed System and its Control Processes' in Spence, K. & Spence, J. (eds), *The Psychology of Learning & Motivation,* New York: Academic Press.

Aylward, E.H. & Burt, D.B. (1998), *'Test Battery for the Diagnosis of Dementia in Individuals with Intellectual Disability. Report of the Working Group for the Establishment of Criteria for the Diagnosis of Dementia in Individuals with Intellectual Disability',* Washington, DC: American Association on Mental Retardation.

APA - American Psychiatric Association (1994*), 'Diagnostic and Statistical Manual of Mental Disorders (DSM-IV). 4th ed',* Washington DC: American Psychiatric Association, 139-143.

Arendt, T. (2001), 'Disturbance of neuronal plasticity is a critical pathologic event in Alzheimer's disease', *International Journal of*

Developmental Neuroscience, 19: 231-245.

Aylward, E.H., Burt, D.B., Thorpe, L.U., Lai, F. & Dalton, A.J. (1997), ‘Diagnosis of dementia in individuals with intellectual disability’, *Journal of Intellectual Disability Research, 41:* 152-164.

Aylward, E.H., Li, Q. & Habbak, R. (1997), ‘Basal ganglia volume in adults with Down’s syndrome’, *Psychiatric Research: Neuroimaging Section, 74:* 73-82.

Baddeley, A.D. (1978), 'The trouble with levels: a re-examination of Craik and Lockhart's framework for memory research', *Psychological Review, 85;* 139-152.

Baddeley, A.D. (1986), *‘Working Memory’*, Oxford: Oxford University Press.

Baddeley, A.D. (1992), 'Working memory', *Science, 255:* 556-559.

Baddeley, A.D. (1996), *‘Human Memory, Rev. ed’*, Hove: Psychology Press.

Baddeley, A.D. (1998), ‘The central executive: A concept and some misconceptions’, *Journal of the International Neuropsychological Society, 4:* 523-526.

Baddeley, A.D., Bressi, S., Della Sala, S., Logie, R. (1991), 'The decline of working memory in Alzheimer's disease', *Brain, 114;* 2521-2542.

Baddeley, A.D. & Hitch, G.J. (1974), 'Working Memory' in Bower, G. (ed*), Recent Advances in Learning and Motivation. Volume III*, London: Academic Press.

Baddeley, A.D., Logie, R., Bressi, S. & Della Sala, S. (1986), 'Dementia and working memory', *Quarterly Journal of Experimental Psychology, 38A;* 603-618.

Baddeley, A.D., Wilson, B.A. & Watts, F. (1995), *'Handbook of Memory Disorders'*, Chichester: Wiley.

Balota, D.A., Tse, C-S., Hutchison, K.A., Spieler, D.H., Duchek, J.M. & Morris, J.C. (2010), 'Predicting conversion to dementia of the Azheimer's type in a healthy control sample: The power of errors in Stroop color naming', *Psychology & Aging, 25,* 1: 208-218.

Barr, O. (1990), 'Down's syndrome and Alzheimer's disease - what is the link?' *Professional Nurse, June;* 465-468.

Bayer, T.A., Wirths, O. & Majtényl, K. (2001), 'Key factors in Alzheimer's disease: β-amyloid precursor protein processing, metabolism and intraneuronal transport', *Brain Pathology, 11:* 1-11.

Becker, J.T. (1988), 'Working memory and secondary memory deficits in Alzheimer's disease', *Journal of Clinical & Experimental Neuropsychology, 6;* 739-753.

Benedict, K.B. & Nacoste, D.B. (1990), 'Dementia and depression in the elderly: A framework for addressing difficulties in differential diagnosis', *Clinical Psychology Review, 10;* 513-537.

Berardi, A., Gaillard, F., Haxby, J.V., Greenwood, P. & Parasuraman, R. (1992), *'Sustained attention in dementia of the Alzheimer type. Paper presented at the Annual Meeting of the Gerontological Society of America'*, Washington DC.

Bigler, E.D. (1992), 'The neurobiology and neuropsychology of adult learning disorders', *Journal of Learning Disability, 25;* 8: 488-506.

Blackburn, I-M. & Davidson, K. (1990), *'Cognitive Therapy for Depression and Anxiety: A Practitioner's Guide, 1st reprint'*, Oxford: Blackwell.

Braekus, A. (1995), 'A low, 'normal' score on the Mini-Mental State Examination predicts development of dementia after three years', *Journal of The American Geriatrics Society, 46:* 656-661.

Beatty, W.W., Rogers, C.L., Rogers, R.L., English, S., Testa, J.A., Orbelo, D.M., Wilson, D.A. & Ross, E.D. (1999), 'Piano playing in Alzheimer's disease: Longitudinal study of a single case.' *Neurocase, 5:* 459-469.

Briggs, A. (1972), *'Report of the Committee on Nursing'*, London:

HMSO.

Burack, O.R. & Lachman, M.E. (1996), 'The effects of list-making on recall in young and elderly adults', *Journal of Gerontology, 51;* 4: P226-P33.

Burns, A., Bernabei, R., Bullock, R., Cruz Jentoft, A.J., Frölich, L., Hock, C., Raivio, M., Triau, E., Vandewoude, M., Wimo, A., Came, E., van Baelen, B., Hammond, G.L., van Oene, J.C. & Schwalen, S. (2009), 'Safety and efficacy of galantamine (Reminyl) in severe Alzheimer's disease (the SERAD study): A randomised, placebo-controlled, double-blind trial', *The Lancet Neurology, 8;* 1: 22-23.

Burns, A.B. & Levy, R. (1994), *'Dementia'*, London: Chapman & Hall.

Burt, D.B. & Aylward, E.H. (2000), 'Test battery for the diagnosis of dementia in individuals with intellectual disability', *Journal of Intellectual Disability Research, 44;* 2: 175-180.

Burt, D.B., Primeaux-Hart, S., Phillips, N.B., Greene, T., Loveland, K.A., Cummings, E., Lewis, K.R., Lesser, J., Cleveland, L. & Chen, Y.R. (1999), 'Assessment of orientation. Relationship between informant report and direct measures', *Mental Retardation, 37:* 364-370.

Chapman, P.F., Falinska, A.M., Knevett, S.G. (2001), 'Genes, models and Alzheimer's disease', *Trends in Genetics, 17;* 5: 254-261.

Chui, H.C., Victoroff, J.I., Margolin, D., Jagust, W., Shankle, R. & Katzman, R. (1992), ‘Criteria for the diagnosis of ischaemic vascular dementia proposed by the State of California Alzheimer’s Disease Diagnostic and Treatment Centers’, *Neurology, 42:* 473-480.

Clare, L., Whitaker, C.J. & Nelis, S.M. (2010), ‘Appraisal of memory functioning and memory performance in healthy ageing and early-stage Alzheimer’s disease’, *Aging, Neuropsychology & Cognition, 18 Mar,* doi: 10.1080/13825580903581558, Accessed: 09.04.2010.

Cohen, R.A. & Albers, H.E. 1991), ‘Disruption of human circadian and cognitive regulation following a discrete hypothalamic lesion: a case study’’ *Neurology, 41:* 726-729.

Cohen, N.J. & Squire, L.R. (1981), 'Preserved learning and retention of pattern analysing skill in amnesia: dissociation of knowing how and knowing that', *Science, 210;* 207-209.

Collacott, R.A. (1992), 'The effect of age and residential placement on adaptive behaviour of adults with Down's syndrome', *British Journal of Psychiatry, 161;* 675-679.

Collerton, D. (1993), 'Memory Disorders' in Greenwood, R., Barnes, M.P., McMillan, T.M. & Ward, C.D. (eds), *Neurological Rehabilitation*, Edinburgh: Churchill Livingstone.

Cooper, J.D., Salehi, A., Delcroix, J-D., Howe, C.L., Belichenko, P.V., Chua-Couzens, J.C., Kilbridge, J.F., Carlson, E.J., Epstein, C.J. & Mobley, W.C. (2001), ‘Failed retrograde transport of NGF in a mouse model of of down’s syndrome: Reversal of cholinergic neurodegenrative phenotypes following NGF infusion’, *Neurobiology, 98;* 18: 10439-10444.

Cooper, P.N., Jackson, M., Lennox, G., *et al.* (1995), ‘Tau, ubiquitin, and alpha β-crystallin immunohistochemistry define the principal causes of degenerative frontotemporal dementia’, *Archives of Neurology, 52:* 1011.

Cooper, S-A. & Collacott, R.A. (1993), 'Prognosis of depression in Down's syndrome', *Journal of Nervous & Mental Disease, 181;* 204-205.

Corder, E.H., Saunders, A.M., Strittmatter, W.J. (1993), ‘Gene dose of apolipoprotein E type 4 allele and the risk of Alzheimer’s disease in late onset families’, *Science, 261:* 921-923.

Corsellis, J.A.N. & Evans, P.H. (1965), 'The Relation of Stenosis of the Extracranial Cerebral Arteries to Mental Disorder and Cerebral Degeneration in Old Age' in Lühy, F. & Bischoff, A. (eds*), Proceedings of the Vth International Congress of Neuropathology*,

Amsterdam: Excerpta Medica, 546-548.

Cox, Y. (1993), 'Tailor-made job', Nursing Times, 89; 22: 66.

Craik, F.I.M. (1994), 'Memory changes in normal aging', Current Directions in Psychological Science, 3; 5: 155-158.

Cullen, C. (1991), *'Mental Handicap Nursing in the Context of 'Caring for People''*, London: Department of Health.

Cummings, J.L. (1992), 'Subcortical dementia as a manifestation of cerebrovascular disease. Proceedings of the NINDS-AIREN International Workshop on Vascular Dementia. NIH, Bethesda, MD, April 19-21, 1991', *New Issues in Neurosciences, 4:* 151-158.

Curran, S. & Wattis, J.P. (1989), 'Round-up: Searching for the cause of Alzheimer's disease', *Geriatric Medicine, March;* 13-14.

Daigneault, S. & Braun, C.M. (1993), 'Working memory and the self-ordered pointing task: further evidence of early prefrontal decline in normal aging', *Journal of Clinical and Experimental Neuropsychology, 15;* 6: 881-895.

Das, J.P. & Jarman, R. (1991), 'Cognitive integration: Alternative model for intelligence' in Rowe, H.A.H. (ed), *Intelligence: Reconceptualization and Measurement*, Hillsdale, New Jersey: Lawrence Erlbaum, 20-21.

Das, J.P., Davison, M., Hiscox, M., Mishra, R.K. (1993), *'Intellectual decline and ageing: Individuals with Down syndrome compared to other individuals with mental handicaps'*, Report, Edmonton: University of Alberta.

Das, J.P., Mishra, R.K., Davison, M. & Naglieri, J.A. (1995), 'Measurement of dementia in individuals with mental retardation', *Clinical Neuropsychologist, 9;* 1: 32-37.

Davis, C., Heidler-Gary, J., Gottesman, R.F., Crnion, J., Newhart, M., Moghekar, A., Soloman, D., Rigamonti, D., Cloutman, L. & Hillis, A.E. (2010), 'Action versus animal naming fluency in subcortical dementia, frontal dementias, and Alzheimer's disease', *Neurocase, 6:* 1-8.

De Groot, J.C., de Leeuw, F-E., Oudkerk, M., Hofman, A., Jolles, J. & Breteler, M.M.B. (2000a), 'Cerebral white matter lesions and depressive symptoms in elderly adults', *Archives of General Psychiatry, 57:* 1071-1076.

De Groot, J.C., de Leeuw, F-E., Oudkerk, M., van Gijn, J., Hofman, A., Jolles, J. & Breteler, M.M.B. (2000b), 'Cerebral white matter lesions and cognitive function: The Rotterdam Scan Study', *Annals of Neurology, 47:* 145-151.

Deb, S. (1997), 'Structural neuroimaging in learning disability'. *British Journal of Psychiatry, 171:* 417-419.

Deb, S. & Braganza, J. (1999), 'Comparison of rating scales for the diagnosis of dementia in adults with Down's syndrome', *Journal of Intellectual Disability Research, 43 (Part 5):* 400-407.

DeKosky, S.T. (2001), 'Epidemiology and pathophysiology of Alzheimer's disease', *Clinical Cornerstone, 3:* 4: 15-24.

Devinsky, O., Sato, S., Conwit, R.A. & Schapiro, M.B. (1990), 'Relation of EEG, alpha background to cognitive function, brain atrophy, and cerebral metabolism in Down's syndrome. Age-specific changes', *Archives of Neurology, 47;* 1: 58-62.

Dickson, D.W. (1997), 'Pathogenesis of senile plaques', *Journal of Neuropathology & Experimental Neurology, 56:* 321.

Dickson, D.W. (1998), 'Astrocytes in Other Neurodegenerative Diseases' in Schipper, H.M. (ed), *Astrocytes in Brain Aging and Neurodegeneration*, RG Landes, Austin, TX, 165.

Dickson, D.W. (2001), 'Neuropathology of Alzheimer's disease and other dementias', *Clinics in Geriatric Medicine, 17;* 2: 209-228.

Dickson, D.W., Crystal, H.A. & Bevona, C. (1995), 'Correlations of synaptic and pathological markers with cognition of the elderly',

Neurobiology & Aging, 16: 285.

Down, J.L. (1866), 'Observation on an ethnic classification of idiots', *London Hospital Reports, 3:* 259-262.

Dubois, B., Feldman, H., Jacova, C., DeKosky, S., Barberger-Gateau, J., Cummings, A., Delacourte, D., Galasko, D., Gauthier, S. & Jicha, G. (2007), 'Research criteria for the diagnosis of Alzheimer's disease: Revising the NINCDS-ADRDA criteria', *The Lancet Neurology, 6;* 8: 734-746.

Dudek, F.J. (1979), 'The continuing misinterpretation of the standard error of measurement', *Psychological Bulletin, 86;* 335-337.

Earles, J.L. & Salthouse, T.A. (1995), 'Interrelations of age, health, and speed', *Journal of Gerontology, 50;* 1: P33-P41.

Eich, E. (1984), 'Memory for unattended events; remembering with and without awareness', *Memory & Cognition, 12:* 105-111.

Ellis, R.D., Goldberg, J.H. & Detweiler, M.C. (1996), 'Predicting age-related differences in visual information processing using two-stage queing model', *Journal of Gerontology, 51;* 3: P155-P65.

Emerling, F. (1990), 'An investigation of test bias in two non-verbal cognitive measures for two ethnic groups', *Journal of Psychoeducational Assessment, 8;* 1: 34-41.

Engelborghs, S. & De Deyn, P.P. (2001), 'Biological and genetic markers of sporadic Alzheimer's disease', *Acta Medica Okayama, 55;* 2: 55-63.

Evenhuis, H.M. (1990), 'Evaluation of a standardized questionnaire as an instrument for diagnosis of dementia in Down's syndrome' in *Clinical Studies of Alzheimer's Dementia and Hearing Loss in Down's Syndrome*, PhD Thesis, University of Amsterdam, Amsterdam.

Evenhuis, H.M. (1992a), *'Provisional Manual of the Dementia Questionnaire for Mentally Retarded Persons (DMR)'*, Zwammerdam: Hooge Burch.

Evenhuis, H.M. (1992b), 'Evaluation of a screening instrument for dementia in ageing mentally retarded persons', *Journal of Intellectual Disability Research, 36:* 337-347.

Evenhuis, H.M. (1996), 'Further evaluation of the Dementia Questionnaire for Persons with Mental Retardation (DMR)', *Journal of Intellectual Disability Research, 40;* 4: 369-373.

Evenhuis, H.M. (1997), 'The natural history of dementia in ageing people with intellectual disability', *Intellectual Disability Research, 41(Pt 1):* 92-96.

Evenhuis, H.M., Eurlings, H.A.L. & Kengen, M.M.F. (1984),

'Diagnostiek van dementie bijbejaarde zwakzinnigen', ['Diagnosis of dementia in mentally retarded persons'], *Ruit, 40:* 14-24.

Evenhuis, H.M., Kengen, M.M.F. & Eurlings, H.A.L. (1990), *'Dementie Vragenlijst Zwakzinnigen', ['Dementia Questionnaire for Mentally Retarded Persons'],* Zwammerdam: Hooge Burch.

Eyman, R.K., Call, T.L. & White, J.F. (1991), 'Life expectancy of persons with Down's syndrome', *American Journal of Mental Retardation, 95:* 603-612.

Filoteo, J.V., Delis, D.C., Massman, P.J., Demadura, T., Butters, N. & Salmon, D.P. (1992), 'Directed and divided attention in Alzheimer's disease: Impairment in shifting attention to global and local stimuli', *Journal of Clinical & Experimental Neuropsychology, 14:* 871-883.

Fisk, A.D., Rogers, W.A., Cooper, B.P. & Gilbert, D.K. (1997), 'Automatic category search and its transfer: aging, type of search, and level of learning', *Journal of Gerontology, 52;* 2: P91-P102.

Flaten, T.P. (2001), 'Aluminium as a risk factor in Alzheimer's disease, with emphasis on drinking water', *Brain research bulletin, 55;* 2: 187-196.

Folstein, M., Folstein, S. & McHugh, P.R. (1975), 'Mini-mental state; a practical method for grading the cognitive state of patients for the

clinician', *Journal of Psychiatric Research, 12:* 189-198.

Freidl, W., Schmidt, R., Stronegger, W.J. & Reinhart, B. (1997), 'The impact of sociodemographic, environmental, and behavioural factors, and cerebrovascular risk factors as potential predictors of the Mattis Dementia Rating Scale', *Journal of Gerontology, 52;* 2: M111-M116.

Fullerton, S., Clark, A., Weiss, K. (2000), 'Apolipoprotein E variation at the sequence haplotype level: Implications for the origin and maintenance of a major human polymorphism', *American Journal of Human Genetics, 67:* 881-900.

Furster, J.M. (1993), 'Frontal lobes', Current Opinions in Neurobiology, 3; 160-165.

Gainotti, G., Parlato, V., Monteleone, D. & Carlomagna, S. (1992), 'Neuropsychological markers of dementia on visuo-spatial tasks: A comparison', *Journal of Clinical & Experimental Neuropsychology, 14;* 2: 239-252.

Gedye, A. (1995), '*Manual for the Dementia Scale for Down's Syndrome'*, Vancouver: Gedye Research and Consulting.

Ghali, L.M. (1996), 'Rest-activity pattern changes in Alzheimer's disease', *Dissertation Abstracts International: Section B; The Sciences & Engineering, 55(1-B):* 6727.

Goate, A., Chartier-Harlin, M.C., Mullan, M. (1991), ‘Segregation of a missense mutation in the amyloid precursor protein gene with familial Alzheimer’s disease’, *Nature, 349:* 704-706.

Goldberg, D. (1972), *‘The Detection of Psychiatric Illness by Questionnaire’*, London: Oxford University Press.

Goldberg, D. & Hillier, V.F. (1979), 'A scaled version of the General Health *Questionnaire', Psychological Medicine, 9:* 139-146.

Goldblum, M-C., Gomez, C-M. & Dalla Barba, G. (1998), ‘The influence of semantic and perceptual encoding on recognition memory in Alzheimer’s disease’, *Neuropsychologia, 36;* 8: 717-729.

Golding, E. (1989), *‘The Middlesex Elderly Assessment of Mental State’*, Bury St Edmunds: Thames Valley Test Company.

Golomb, J., Kluger, A., de Leon, M.J. & Ferris, S.H. (1996), 'Hippocampal formation size predicts declining memory performance in normal aging', *Neurology, 47;* 3: 810-813.

Goudie, F. (1993), 'Problems of Aging' in Stokes, G. & Goudie, F. (eds*), Working with Dementia*, Oxford: Winslow Press, 29-43.

Grafman, J. (1992), ‘Neuropsychological assessment of vascular dementia. Proceedings of the NINDS-AIREN International Workshop on Vascular Dementia. NIH, Bethesda, MD, April 19-21,

1991', *New Issues in Neurosciences, 4:* 159-163.

Grimaldi, L., Cassadei, V. & Ferri, C. (2000), 'Association of early onse Alzheimer's disease with an interleukin-1 alpha gene polymorhism', *Annals of Neurology, 47:* 361-365.

Greene, J.D.W., Hodges, J.R. & Baddeley, A.D. (1995), 'Autobiographical memory and executive function in early dementia of Alzheimer type', *Neuropsychologia, 33;* 12: 1647-1670.

Hagberg, B. & Gustafson, L. (1985), 'On diagnosis of dementia: Psychometric investigation and clinical psychiatric evaluation in relation to verified diagnosis', *Archives of Gerontology & Geriatrics, 4:* 321-332.

Hall, R. & Davis, T.E. (2010), 'Factor structure of the Geriatric Depression Scale in cognitively impaired older adults', *Clinical Gerontologist, 33;* 1: 39-48.

Hanley, I. & Baikie, E. (1984), 'Understanding and treating depression in the elderly' in Hanley, I. & Hodge, J. (eds), *Psychological Approaches to the Care of the Elderly*, Beckenham: Croom Helm.

Heston, L.L., Mastri, A.R., Anderson, V.E. & White, J. (1981), 'Dementia of the Alzheimer type. Clinical genetics, natural history and associated conditions', *Archives of General Psychiatry, 38:* 1085-

1090.

Hofman, A., Ott, A., Breteler, M.M., *et al.* (1997), 'Atherosclerosis, apolipoprotein E, and the prevalence of dementia and Alzheimer's disease in a population-based study: The Rotterdam Study', *Lancet, 349:* 151-154.

Holden, U. (1989), *'Neuropsychology and Ageing'*, London: Chapman & Hall.

Hon, J., Huppert, F.A., Holland, A.J. & Watson, P. (1999), 'Neuropsychological assessment of older adults with Down's syndrome: an epidemiological study using the Cambridge Cognitive Examination (CAMCOG)', *British Journal of Clinical Psychology, 38 (Part 2):* 155-165.

Hopper, T., Drefs, S., Bayles, K.A., Tomoeda, C.K. & Dinu, I. (2009), 'The effects of modified spaced-retrieval training on learning and retention of face-name associations by individuals with dementia', *Neuropsychological Rehabilitation, 20;* 1: 81-102.

HSS - Hampshire Social Services (1989), *'Hampshire Social Services Assessment'*, Southampton: Hampshire Social Services.

Hu, X., Crick, S.L., Bu, G., Freiden, C., Pappu, R.V. & Lee, J-M. (2010), 'Amyloid seeds formed by cellular uptake, concentration, and

aggregation of the amyloid-beta peptide', *Proceedings of the National Academy of Sciences of the United States, 106;* 48: 20324-20329.

Hulstaert, F., Blennow, K., Schoonderwaldt, H.C. (1999), 'Improved discrimination of AD patients using β-amyloid $_{(1\text{-}42)}$ and tau levels in CSF', *Neurology, 52:* 1555-1562.

Hurley, A.C., Volicer, B.J., Hanrahan, P.A., Houde, S. & Volicer, L. (2007), 'Assessment of discomfort in advanced Alzheimer patients', *Research in Nursing & Health, 15;* 5: 369-377.

Isles, A.R. & Wilkinson, L.S. (2000), 'Imprinted genes, cognition and behaviour', *Trends in Cognitive Science, 4;* 8: 309-318.

Jancar, J. (1984), 'Protracted survival in patients with Down's syndrome', *British Medical Journal, 288:* 152.

Jay, P. (1979), *'Report of the Committee of Enquiry into Mental Handicap Nursing and Care'*, London: HMSO.

Jervis, G.A. (1948), 'Early senile dementia in mongoloid idiocy', *American Journal of Psychiatry, 105:* 102-106.

Jorm, A.F. (1990), *'The Epidemiology of Alzheimer's Disease and Related Disorders'*, London: Chapman & Hall.

Kakinuma, Y., Furihata, M, Akiyama, T., Arikawa, M., Handa, T.,

Katare, R.G. & Stao, T. (2010), ‘Donepezil, an acetylcholine esterase inhibitor against Alzheimer’s dementia, promotes angiogenesis in an ischaemic hindlimb model’, *Journal of Molecular & Cellular Cardiology, 48;* 4: 680-693.

Katzman, R. (1976), 'The prevalence and malignancy of Alzheimer's disease', *Archives of Neurology, 33:* 217-8.

Kaufer, D. (1998), ‘Beyond the cholinergic hypothesis: the effect of metrifonate and other cholinesterase inhibitors on neuropsychiatric symptoms in Alzheimer’s disease’, *Dementia & Geriatric Cognitive Disorders, 9;* Suppl 2: 8-14.

Kay, B. (1993), 'Keeping it in the family', *Nursing Times, 89;* 22: 64-65.

Kearns, N.P., Cruikshank, C.A., McGuigan, K., Riley, S.A. (1982), 'A comparison of depression rating scales', *British Journal of Psychiatry, 141:* 45-49.

Kemper, S., LaBarge, E., Farraro, R., Cheung, H., Cheung, H. & Storandt, M. (1993), ‘On the preservation of syntax in Alzheimer’s disease’, *Archives of Neurology, 50:* 81-86.

Kendrick, D. (1985), *‘Kendrick Cognitive Tests for the Elderly. Manual’*, Windsor: NFER-NELSON.

Kengen, M.M.F., Eurlings, H.A.L., Evenhuis, H.M. & de Boer, J. (1987),

'De Dementie Vragenlijst voor Zwakzinnigen (DVZ): Een onderzoeksinstrument voor de diagnostiek van seniele dementie bij zwakzinnigen', ['Dementia Questionnaire for Mentally Retarded Persons (DMR): An instrument for the diagnosis of senile dementia in mentally retarded persons'], *Ruit, 43:* 24-30.

Kesslak, J.P., Nagata, S.F. & Lott, I. (1994), 'Magnetic resonance imaging analysis of age-related changes in the nrain of individuals with Down's syndrome', *Neurology, 44:* 1039-1045.

Kline, J., Kinney, A., Levin, B. & Warburton, D. (2000), 'Trisonomic pregnancy and earlier age at menopause', *American Journal of Human Genetics, 67:* 395-404.

Kopelman, M.D., Wilson, B.A. & Baddeley, A.D. (1990), *'The Autobiographical Memory Interview'*, Bury St Edmunds: Thames Valley Test Co.

Krishnan, K.R. (1991), 'Organic bases of depression in the elderlt', *Annual Review in Medicine, 42:* 261-266.

Krishnan, K.R., Hays, J.C. & Blazer, D.G. (1997), 'MRI-defined vascular depression', *American Journal of Psychiatry, 154:* 497-501.

Krishnan, K.R. & McDonald, W.M. (1995), 'Arteriosclerotic depression', *Medical Hypotheses, 44:* 111-115.

Kumar, V., Anand, R., Messina, J., Hartman, R. & Veach, J. (2008), ‚An efficacy and safety analysis of Exelon in Alzheimer’s disease patients with concurrent vascular risk factors’, *European Journal of Neurology, 7;* 2: 159-169.

LaBerge, D. (1990), ‘Thalamic and cortical mechanisms of attention suggested by recent positron emission tomographic experiments’, *Journal of Neuroscience, 2:* 358-372.

Larsson, T., Sjögren, T. & Jacobson, G. (1963), 'Senile dementia: A clinical, sociomedical and genetic study', *Acta Psychiatrica Scandinavica, 39;* Suppl.: 167.

Leckliter, I.N., Matarazzo, J.D. & Silverstein, A.B. (1986), 'A literature review of factor analytic studies of the WAIS-R', *Journal of Clinical Psychology, 42:* 332-342.

Lekeu, F., Magis, D., Marique, P., Delbeuck, X., Bechet, S., Guillaume, B., Adam, S., Petermans, J., Moonen, G. & Salmon, E. (2009), ‘The California Verbal Learning Test and other standard clinical neuropsychological tests to predict conversion from mild memory impairment to dementia’, *Journal of Clinical & Experimental Neuropsychology, 32;* 2: 164-173.

Levy-Lahad, E., Wasco, W., Poorkaj, P. (1995), ‘Candidate gene for the

chromosme 1 familial Alzheimer's disease locus', *Science, 269:* 973-977.

Lezak, M.D. (1983), *'Neuropsychological Assessment, 2nd ed'*, New York: Oxford University Press.

Li, D., Hu, K., Chen, G., Jin, Y. & Teng, E.L. (1988), 'Combined form of Raven's Test (CRT) and its preliminary testing in the Shanghai Metropolitan Area', *Correspondence of the Psychological Sciences Journal, 4:* 27-31.

Lines, C.R., Dawson, C., Preston, G.C., Reich, S., Foster, C. & Traub, M. (1991), 'Memory and attention in patients with senile dementia of the Alzheimer type and normal elderly subjects', *Journal of Clinical & Experimental Neuropsychology, 13:* 691-702.

Lishman, W.A. (1987), *'Organic Psychiatry: the Psychological Consequences of Cerebral Disorder, 2nd ed'*, Oxford: Blackwell.

Lord, F.M. & Novick, M.R. (1968), *'Statistical Theories of Mental Test Scores'*, Massachusetts: Addison-Wesley.

Luchsinger, J.A., Tang, M-X. & Stern, Y. (2001), 'Diabetes mellitus and risk of Alzheimer's disease and dementia with stroke in a multi ethnic cohort', *American Journal of Epidemiology, 154;* 7: 635-641.

Maclean, W.E., Ellis, D.N., Galbreath, H.N. & Halpern, L.F. (1991),

'Rhythmic motor behaviour of preambulatory motor impaired, Down syndrome and nondisabled children - A comparative analysis', *Journal of Abnormal Child Psychology, 19:* 319-330.

Mahley, R.W. & Rall, S.C. (2000), 'Apolipoprotein E: Far more than a Lipid Transport Protein', *Annual review of Genomics & Human Genetics, 1:* 507-537.

Marchegiani, A., Giannelli, M.V. & Odetti, P.R. (2009), 'The Tower of London test: A test for dementia', *Aging & Mental Health, 14;* 2: 155-158.

Markesberry, W.R. (1998), 'Vascular Dementia' in Markesberry, W.R. (ed), *Neuropathology of Dementing Disorders,* Arnold, New York, 293.

Matarazzo, J.D. (1972), *'Wechsler's Measurement and Appraisal of Adult Intelligence, 5th ed'*, Baltimore: Williams & Wilkins.

Mattis, S. (1988), *'Dementia Rating Scale'*, Windsor: NFER-NELSON.

McDonald, A.J.D. (1986), 'Do general practitioners "miss" depression in elderly patients?' *British Medical Journal, 292;* May: 1365-1367.

McDowell, I. (2001), 'Alzheimer's disease: Insights from epidemiology', *Aging (Milano), 13;* 3: 143-162.

McNemar, Q. (1957), 'On WAIS difference scores', *Journal of Consulting*

Psychology, 21: 239-240.

Measso, G., Zappala, G., Cavarzeran, F. & Crook, T.H. (1993), 'Raven's Coloured Progressive Matrices: A random study of healthy adults', *Acta Neurologica Scandinavica, 18;* 1: 70-74.

Mendez, M.F., Mastri, A.R. & Sung, J.H. (1992), 'Clinically diagnosed Alzheimer's disease: Neuropathologic findings in 650 cases', *Alzheimer Disease & Associated Disorders, 6:* 35.

Mental Health Foundation (1993), *'Learning Disabilities: The Fundamental Facts'*, London: Mental Health Foundation.

Merched, A., Blain, H. & Visvikis, S. (1997), 'Cerebrospinal fluid apoplipoprotein E level is increased in late-onset Alzheimer's disease', *Journal of the Neurological Sciences, 145:* 33-39.

Miller, E. & Morris, R. (1993), *'The Psychology of Dementia'*, Chichester: John Wiley.

Milne, A. (2010), 'Dementia screening and early diagnosis: The case for and against', *Health, Risk & Society, 12;* 1: 65-76.

Mitchell, K. (1998), 'Assessing Dementia and Depression in Adults with Mental Retardation' in Rotatori, A., Schwenn, J.O. (eds), *Advances in Special Education, Vol. 11: Issues, Practices and Concerns in Special Education*, Jai Press: Greenwich, CT, 255-264.

Mitrushina, M., Uchiyama, C. & Satz, P. (1995), 'Heterogeneity of cognitive profiles in normal aging: implications for early manifestations of Alzheimer's disease', *Journal of Clinical and Experimental Neuropsychology, 17;* 3: 374-382.

Mohanaruban, K., Sastry, B.S.D. & Finucane, P. (1989), 'Assessment and diagnosis of dementia', *Geriatric Medicine, June:* 81-84.

Mohr, E., Cox, C., Williams, J., Chase, T.M., & Fedio, P. (1990), 'Impairment of central auditory function in Alzheimer's disease', *Journal of Clinical & Experimental Neuropsychology, 12:* 235-246.

Money, E.A., Kirk, R.C. & McNaughton, N. (1992), 'Alzheimer's dementia produces a loss of discrimination but no increase in rate of memory decay in delayed matching to sample', *Neuropsychologia, 30:* 133-143.

Moody, P. & Moody, R. (1992), *'Half Left'*, Oslo: Dreyers Forlag A/S.

Morris, J.C. (1999), 'Clinical Presentation and Course of Alzheimer's Disease' in Bick, K.L., *et al.* (eds), *Alzheimer Disease Edition 2,* Lippincott, Williams & Wilkins, Philadelphia, 11.

Morris, R.G. (1984), 'Dementia and the functioning of the articulatory loop system', *Cognitive Neuropsychology, 1:* 143-157.

Morris, R.G. (1986), 'Short-term forgetting in senile dementia of the

Alzheimer's type', *Cognitive Neuropsychology, 3:* 77-97.

Morris, R.G. (1987a), 'The effect of concurrent articulation on memory span in Alzheimer-type dementia', *British Journal of Clinical Psychology, 26:* 233-234.

Morris, R.G. (1987b), 'Articulatory rehearsal in Alzheimer-type dementia', *Brain & Language, 30:* 351-362.

Morris, R.G. (1992), 'Patterns of Short-Term Memory Impairment in Alzheimer's Disease' in Backman, L. (ed), *Memory Functioning in Dementia*, Amsterdam: Elsevier, 3-25.

Morris, R.G. (1994a), 'Working memory in Alzheimer-type dementia', *Neuropsychology, 8;* 4: 544-554.

Morris, R.G. (1994b), 'Recent developments in the neuropsychology of dementia', *International Review of Psychiatry, 6:* 85-107.

Morris, R.G. (ed) (1996), *'The Cognitive Neuropsychology of Alzheimer-Type Dementia'*, Oxford, Oxford University Press.

Moss, S., Goldberg, D. & Patel, P. (1991), *'Psychiatric and physical morbidity in older people with severe mental handicap'*, Report, University of Manchester: Hester Adrian Centre.

Moss, S.C. & Patel, P. (1992), 'Symptoms of Dementia in Older People with Severe Mental Handicap: Cognitive and Functional Decline

Over a Three-Year Period' in Roosendaal, J.J. (ed), *Mental Retardation and Medical Care,* CIP-Gegevens Konklijke Bibliotheek: Den Haag, 289-297.

Moss, S.C. & Patel, P. (1995), 'Psychiatric symptoms associated with dementia in older people with learning disability', *The British Journal of Psychiatry, 167;* 5: 663-667.

Myers, A.J. & Goate, A.M. (2001), 'The genetics of late-onset Alzheimer's disease', *Current Opinion in Neurology, 14;* 433-440.

Myers, A., Holmans, P. & Marshall, H. (2000), 'Susceptibility locus for Alzheimer's disease on chromosome 10', *Science, 290:* 2304-2305.

Nagy, Z., Vatter-Bittner, B., Braak, H., Braak, E., Yilmazer, D.M., Schultz, C. & Hanke, J. (1998), 'Staging of Alzheimer-type pathology: An interater-infrarater study', *Dementia & Geriatric Cognitive Disorders, 8;* 4: 248-251.

Nemes, Z., Fésüs, L., Égerhazi, A., Keszthelyi, A. & Degrell, I.M. (2001), 'N^{ε}(γ-glutamyl) lysine in cerebrospinal fluid marks Alzheimer type and vascular dementia', *Neurobiology of Aging, 22:* 403-406.

Nizetic, D. (2001), 'Functional genomics of the Down syndrome', *Croatian Medical Journal, 42 (4):* 421-427.

Nordberg, A., Rinne, J.O., Kadir, A. & Långström, B. (2010), 'The use of

PET in Alzheimer disease', *Nature Reviews Neurology, 6:* 78-87.

Norman, D.A. & Shallice, T. (1986), 'Reflections on Cognition and Parallel Distributed Processing' in McClelland, J.L. & Rumelhart, D.E. (eds), *Parallel Distributed Processing: Exploration of the Microstructure of Cognition. Volume 2*, Cambridge, Massechusetts: MIT Press, 102-110.

Nyberg, L., Backman, L., Erngrund, K., Olofsson, U. & Nilsson, L.G. (1996), 'Age differences in episodic memory, semantic memory, and priming: relationships to demographic, intellectual, and biological factors', *Journal of Gerontology, 51;* 4: P234-P40.

O'Brien, J., Ames, D., Chui, E. & Schweietzer, I. (1998), 'Severe deep white matter lesions and outcome in elderly patients with major depressive disorder: Follow up study', *British Medical Journal, 317:* 982-984.

O'Brien, J., Desmond, P., Ames, D. & Schweitzer, I. (1996), 'A magnetic resonance imaging study of white matter lesions in depression and Alzheimer's disease', *British Journal of Psychiatry, 168:* 477-485.

Oliver, C. (1999), 'Perspectives on Assessment' in Janicki, M.P., Dalton, A.J. (eds), *Dementia, Aging, and Intellectual Disabilities: A Handbook*, Brunner/Mazel: Philadelphia, PA, 123-140.

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

Oswald, F., Wahl, H-W., Voss, E., Schilling, O., Freytag, T., Auslander, G., Shoval, N., Heinik, J. & Landau, R. (2010), ‚The use of tracking technologies for the analysis of outdoor mobility in the face of dementia: First steps into a project and some illustrative findings from Germany’, *Age & Aging, 24;* 1: 55-73.

Palmer, C.G.S., Crank, C., Pueschel, S.M. & Wisniewski, K.E. (1992), 'Head circumference of children with Down's syndrome (0-36 months)', *American Journal of Medical Genetics, 42:* 61-67.

Paul, R.H., Cohen, R.A., Moser, D. & Ott, B.R. (2001), ‘Performance on the Mattis Dementia Rating Scale in patients with vascular dementia: Relationships to neuroimaging findings’, *Journal of Geriatric Psychiatry & Neurology, 14:* 33-36.

Parasuraman, R. & Giambra, L. (1991), ‘Skill development in vigilance: Effects of event rate and age’, *Psychology & Aging, 6:* 155-169.

Parasuraman, R., Greenwood, P.M., Haxby, J.V. & Grady, C.L. (1992), ‘Visuospatial attention in dementia of tye Alzheimer’s type’, *Brain, 115:* 711-733.

Parkin, A.J., Walter, B.M. & Hunkin, N.M. (1995), 'Relationships

between normal aging, frontal lobe function, and memory for temporal and spatial information', *Neuropsychology, 9;* 3: 304-312.

Pattie, A. & Gilleard, C. (1979), *'Clifton Assessment Procedures for the Elderly'*, Windsor: NFER-NELSON.

Pericak-Vance, M.A., Grubber, J. & Bailey, L.R. (2000), 'Identification of novel genes in late-onset Alzheimer's disease', *Experimental Gerontology, 35:* 1343-1352.

Piedmont, R.L., Sokolove, R.L. & Fleming, M.Z. (1989), 'On WAIS-R difference scores in a psychiatric sample', Psychological Assessment: *A Journal of Consulting & Clinical Psychology, 1:* 155-159.

Pinter, J.D., Eliez, S. & Schmitt, J.E. (2001), 'Neuroanatomy of Down's syndrome: A high-resolution MRI study', *American Journal of Psychiatry, 158;* 10: 1659-1665.

Pollak, C.P., Perlick, D. & Linsner, J.P. (1993), 'Daily sleep reports and circadian rest-activity cycles of elderly community residents with insomnia', *Biological Psychiatry, 32;* 11: 1019-1027.

Potter, H. (1991), 'Review and hypothesis: Alzheimer disease and Down syndrome – chromosome 21 nondisjunction may underlie both disorders', *American Journal of Human Genetics, 48:* 1192-1200.

Powell, T., Brooker, D.J.R. & Papadopolous, A. (1993), 'Test-retest

reliability of the Middlesex Assessment of Mental State (MEAMS): A preliminary investigation in people with probable dementia', *British Journal of Clinical Psychology, 32:* 224-226.

Prasher, V.P., Barber, P.C. & West, R. (1996), 'The role of magnetic resonance imaging in the diagnosis of Alzheimer's disease in adults with Down's syndrome: case report', *Archives of Neurology, 53:* 1310-1313.

Pulsifer, M.B. (1996), 'The neuropsychology of mental retardation', *Journal of the International Neuropsychological Society, 2 (2):* 159-176.

Rao, R. (1998), 'Depression after transient ischaemic attack: A clinically distinct subtype of vascular depression?' *Archives of general Psychiatry, 55:* 753-754.

Robinson, D.M. & Keating, G.M. (2006), 'Memantine: A review of its use in Alzheimer's disease', *Drugs 66*; 11: 1515–1534.

Ruitenberg, A., Kalmijn, S. & de Ridder, M.A. (2001), 'Prognosis of Alzheimer's disease: The Rotterdam study', *Neuroepidemiology, 20;* 3: 188-195.

Ruitenberg, A., Ott, A. & van Swieten, J.C. (2001), 'Incidence of dementia: Does gender make a difference?' *Neurobiology of Aging,*

22: 575-580.

Qizilbash, N., Whitehead, A., Higgins, J., Wilcock, G., Schneider, L. & Farlow, M. (1998), 'Cholinesterase inhibition for Alzheimer disease: a meta-analysis of the tacrine trials. Dementia Trialists Collaboration', *Journal of the American medical Association, 280;* 20: 1777-1782.

Rau, M.T. (1993), *'Coping with Communication Challenges and Alzheimer's'*, London: Chapman & Hall.

Raven, J.C. (1956), *'Guide to Using the Coloured Progressive Matrices, Revised ed'*, Oxford: Oxford Psychologists Press.

Raven, J.C. (1965), *'Guide to Using the Coloured Progressive Matrices, 1st reprint'*, Oxford: Oxford Psychologists Press.

Raven, J.C., Court, J.H. & Raven, J. (1990), *'Raven's Progressive Matrices and Vocabulary Scales. Section 2: Coloured Progressive Matrices'*, Oxford: Oxford Psychologists Press.

Raven, J.C., Court, J.H. & Raven, J. (1995), *'Raven Manual: Section 2. Coloured Progressive Matrices 1995 Edition'*, Oxford: Oxford Psychologists Press.

Reisberg, B. (1983), 'An Overview of Current Concepts of Alzheimer's Disease, Senile Dementia, and Age-Associated Cognitive Decline', in

Reisberg, B. (ed), *Alzheimer's Disease, the Standard Reference*, The Free Press: New York.

Reiss, S. (1987), *'Reiss Screen for Maladaptive Behavior'*, Worthington, OH: International Diagnostic Systems Inc.

Rhodin, J.A. & Thomas, T. (2001), 'A vascular connection to Alzheimer's disease', *Microcirculation, 8;* 4: 207-220.

Román, G.C., Tatemichi, T.K., Erkinjuntti, T., Cummings, J.L., Masdeu, J.C., Garcia, J.H., Amaducci, L., Orgogozo, J-M., *et al.* (1993), 'Vascular dementia: Diagnostic criteria for research studies', *Neurology, 43:* 250-260.

Román, G.C. (1999), 'Vascular dementia today', *Revue Neurologie (Paris), 155:* S64-S72.

Rossor, M.N. (1999), 'Differential diagnosis of frontotemporal dementia: Pick's disease', *Dementia & Geriatric Cognitive Disorders, 10 (Suppl):* 43.

Roth, M. (1955), 'The natural history of mental disorders in old age', *Journal of Mental Science, 102:* 281-293.

Sahakian, B.J., Morris, R.G., Evenden, J.L., Heald, A., Levy, R., Philpot, M. & Robbins, T.W. (1988), 'A comparative study of visuospatial memory and learning in Alzheimer-type dementia and Parkinson's

disease', *Brain, 111:* 698-718.

Salthouse, T.A. & Meinz, E.J. (1995), 'Aging, inhibition, working memory, and speed', *Journal of Gerontology, 50;* 6: P297-P306.

Satlin, A., Volicer, L., Stopa, E.G. & Harper, D. (1996), 'Circadian locomotor activity and core-body temperature rhythms in alzheimer's disease', *Neurobiology of Aging, 16;* 5: 765-771.

Sattel, H., Geiger-Kabisch, C. & Schreiter-Gasser, U. (1993), 'Incidence and importance of 'non-cognitive' symptoms in dementia of the Alzheimer type: productive psychotic symptoms, depressive disorders and behavioral disorders', *Zeitchrift fur Gerontologie, 26:* 275-279.

Saunders, A.M. (2000), 'Apolipoprotein E and Alzheimer's disease: An update of genetic and functional analysis', *Journal of Neuropathology & Experimental Neurology, 59:* 751-758.

Schapiro, M.B., Haxby, J.V. & Grady, C.L. (1992), 'Nature of mental retardation and dementia in Down syndrome: Study with PET, CT, and neuropsychology', *Neurobiology of Aging, 13;* 6: 723-734.

Scherder, E.J.A. (1995), *'Peripheral Nerve Stimulation in Alzheimer's Disease: Effects on Memory and Affective Behaviour'*, CIP-Gegevens Koninklijke Bibliotheek: Den Haag.

Schiavi, R.C. & Rehman, J. (1995), 'Sexuality and aging', *Urology Clinics of North America, 22;* 4: 711-726.

Schmand, B., Geerlings, M.I., Jonker, C. & Lindeboom, J. (1998), ‘Reading ability as an estimator of premorboid intelligence: Does it remain stable in emergent dementia?’ *Journal of Clinical & Experimental Neuropsychology, 20;* 1: 42-51.

Schmitt, A.L., Livingston, R.B., Smernoff, E.N., Reese, E.M., Hafer, D.G. & Harris, J.B. (2010), ‘Factor analysis of the Repeatable Battery for the Assessment of Neuropsychological Status (RBANS) in a large sample of patients suspected o dementia’, *Applied Neuropsychology, 17;* 1: 8-17.

Schneck, M.H., Reisberg, B. & Ferris, S.H. (1982), 'An overview of current concepts of Alzheimer's disease', *American Journal of Psychiatry, 139:* 165-173.

Schupf, N., Kapell, D., Nightingale, B., Lee, J.H., Mohlenhoff, J., Bewley, S., Ottman, R. & Mayeux, R. (2001), ‘Specificity of the fivefold increase in AD in mothers of adults with Down’s syndrome’, *Neurology, 57:* 979-984.

Scott, W.K., Grubber, J.M. & Conneally, P.M. (2000), ‘Fine mapping of the chromosome 12 late-onset Alzheimer disease locus: Potential

genetic and phenotypic heterogeneity', *American Journal of Human Genetics, 66:* 922-932.

Seguin, E. (1866), *'Idiocy: Its Treatment by the Physiological Method'*, New York: William Wood.

Shallice, T. (1988), *'From Neuropsychology to Mental Structure'*, New York: Cambridge University Press.

Shermann, S.L., Takesu, N., Freeman, S.B. & Grantham, M. (1991), 'Association between reduced recombination with nondisjunction', *American Journal of Human Genetics, 49:* 608-620.

Sherrington, R., Rogaev, E.I., Liang, Y. (1995), 'Cloning of a gene bearing missense mutations in aerly-onset familial Alzheimer's disease', *Nature, 375:* 754-760.

Simone, P.M. & Baylis, G.C. (1997), 'The role of attention in a spatial memory task in Alzheimer disease patients', *Alzheimer Disease & Associated Disorders, 11;* 3: 140-152.

Skoog, I., Kalaria, R.N. & Breteler, M.M. (1999), 'Vascular factors and Alzheimer's disease', *Alzheimer Disease and Associated Disorders, Suppl 3:* S104-S114.

Small, G.W., La Rue, A., Komo, S., Kaplan, A. & Mandelkern, M.A. (1995), 'Predictors of cognitive change in middle-aged and older

adults with memory loss', *American Journal of Psychiatry, 152;* 12: 1757-1764.

Somerset County Council (1992), *'Care in the Community: Strategy for Persons in Mental Handicap Hospitals'*, Taunton: Somerset County Council.

Spinnler, H. & Della Sala, S. (1988), 'The role of clinical neuropsychology in the neurological diagnosis of Alzheimer's disease', *Journal of Neurology, 235:* 258-271.

Spinnler, H., Della Sala, S., Bandera, R. & Baddeley, A. (1988), 'Dementia, ageing, and the structure of human memory', *Cognitive Neuropsychology, 5:* 193-211.

Stokes, G. & Holden, U. (1993), 'Dementia: Causes & Clinical Syndromes' in Stokes, G. & Goudie, F. (eds), *Working with Dementia*, Bicester: Winslow Press.

Sutton, J.P. & Hobson, J.A. (1998), 'Waking and Sleeping States' in Parks, R.W. & Levine, D. S. (eds), *Fundamentals of Neural Network Modeling: Neuropsychology and Cognitive Neuroscience,* The MIT Press: Cambridge, MA.

Swaab, D.F., Hofman, M.A., Lucassen, P.J., Purba, J.S., Raadsheer, F.C. & van de Nes, J.A.P. (1993), 'Functional neuroanatomy and

neuropathology of the human hypothalamus', *Anatomy & Embyology, 187:* 317-330.

Swearer, J.M. & Kane, K.J. (1996), 'Behavioral slowing with age: boundary conditions of the generalized slowing model', *Journal of Gerontology, 51:* 4: P189-P200.

Tatemichi, T.K., Desmond, D.W. & Paitz, M. (1991), 'The "Mini-Mental State" examination as a screen for dementia following stroke', *Journal of Clinical & Experimental Neuropsychology, 13:* 419.

Terr, L. (1994), *'Unchained Memories: True Stories of Traumatic Memories, Lost and Found'*, London: Basic Books.

Tierney, M.C., Black, S.E. & Szalai, J.P. (2001), 'recognition memory and verbal fluency differentiate probable Alzheimer disease from subcortical ischaemic vascular dementia', *Archives of Neurology, 58;* 10: 1654-1659.

Thompson, S.B.N. (1989), Techniques for tackiling anxiety', *Therapy Weekly, 15;* 49: 6.

Thompson, S.B.N. (1993a), 'Down's syndrome and Alzheimer's disease', Therapy Weekly, 19; 21: 8.

Thompson, S.B.N. (1993b), *'Eating Disorders: A Guide for Health Professionals'*, London: Chapman & Hall.

Thompson, S.B.N. (1994), 'Sexuality training in occupational therapy for people with learning disability, four years on: Policy guidelines', *British Journal of Occupational Therapy, 57;* 7: 255-258.

Thompson, S.B.N. (1995), 'Storage problems', *Therapy Weekly, 21;* 40: 7.

Thompson, S.B.N. (1996), 'Practical ways of improving memory storage and retrieval problems in patients with head injuries', *British Journal of Occupational Therapy, 59;* 9: 418-422.

Thompson, S.B.N. (1997), '*Dementia: A Guide for Health Care Professionals'*. Aldershot: Arena.

Thompson, S.B.N. (1999a), 'Rehabilitation of Cognitive and Emotional Problems', in Fawcus, R. (ed), *Stroke Rehabilitation: A Collaborative Approach,* Oxford, Blackwell.

Thompson, S.B.N. (1999b), 'Assessing dementia in people with learning disabilities for cognitive rehabilitation', *Journal of Cognitive Rehabilitation, 17;* 3: 14-20.

Thompson, S.B.N. (1999c), 'Examining dementia in Down's syndrome (DS): Decline in social abilities in DS compared with other learning disabilities', *Clinical Gerontologist, 20;* 3: 23-44.

Thompson, S.B.N. (2000), 'Investigation into Down's syndrome and dementia', *Journal of the Association of Practitioners in Learning*

Disability, 17; 3: 10-14.

Thompson, S.B.N. (2001a), ‘‘Hollywood wives’: Education and support group for spouses of inpatients with dementia’, *Journal of Dementia Care, 9;* 2: 38.

Thompson, S.B.N. (2001b), *Memory Problems: A Self-Help Guide for Patients, Carers, Health Care Professionals and Students in Training,* Chichester, Oxford Press.

Thompson, S.B.N. & Morgan, M. (1996), *‘Occupational Therapy for Stroke Rehabilitation, 2nd reprint’*. London: Chapman & Hall.

Thompson, S.B.N., North, N. & Pentland, B. (1992), 'Clinical management of a man with complex partial seizures and a severe head injury', *Brain Injury, 7;* 3: 257-262.

Tomlinson, B.E., Blessed, G. & Roth, M. (1970), 'Observations on the brains of demented old people', *Journal of the Neurological Sciences, 11:* 205-242.

Tulving, E. (1972), ‘Episodic and Semantic Memory’ in Tulving, E. & Donaldson, W. (eds), *Organisation of Memory,* New York: Academic Press.

Turnbull, J. (1993), 'Diverse options', *Nursing Times, 89;* 22: 62-63.

Tyrrell, J.F., Cosgrave, M.P., McLaughlin, M. & Lawlor, B.A. (1996),

‘Dementia in an Irish population of Down’s syndrome people’, *Irish Journal of Psychological Medicine, 13;* 2: 51-54.

van Belle, G., Uhlmann, R.F., Hughes, J.P. & Larson. E.B. (1990), 'Reliability of estimates of change in mental status test performance in senile dementia of the Alzheimer type', *Journal of Clinical Epidemiology, 43;* 6: 589-595.

van den Broek, M.D. & Bradshaw, C.M. (1994), 'Detection of acquired deficits in general intelligence using the National Adult Reading Test and Raven's Standard Progressive Matrices', *British Journal of Clinical Psychology, 33:* 509-515.

Van Someren, E.J.W., Kessler, A., Majid, M. & Swaab, D.F. (1997), ‘Indirect bright light improves circadian rest-activity disturbances in demented patients’, *Biological Psychiatry, 41;* 9: 955-963.

Van Someren, E.J.W., Lijzenga, C., Majid, M. & Swaab, D.F. (1998b), ‘Long-term fitness training improves the circadian rest-activity rhythm in healthy elderly males’, *Journal of Biological Rhythms, 12;* 2: 146-156.

Van Someren, E.J.W., Scherder, E.J.A. & Swaab D.F. (1998a), ‘Transcutaneous electrical nerve stimulation (TENS) improves circadian rhythm disturbances in Alzheimer’s disease’, *Alzheimer*

Disease & Associated Disorders, 12; 2: 114-118.

Verhaart, W.J.C. & Jelgersma, H.C. (1952), 'Early senile dementia in mongolian idiocy. Description of a case', *Folia Psychiatrica Neerlandica, 55:* 453-459.

Vincent, L.C. (1996), 'Neuropsychological assessment of developmental learning disabilities in adults', *Seminars in Speech & Language, 17 (3):* 183-200.

Vodegel-Matzen, L.B.L., van der Molen, M.W. & Dudink, A.C.M. (1994), Error analysis of Raven test performance, *Personality & Individual Differences, 16;* 3: 433-445.

Wall, M. (2010), 'The effects of music therapy for older people with dementia', *British Journal of Nursing, 19;* 2: 108-113.

Wang, J., Kwon, J. & Shah, P. (2000), 'Effect of APOE genotype and prometen polymorphism on risk of Alzheimer's disease', *Neurology, 55:* 1644-1649.

Warrington, E.K. (1984), *'Recognition Memory Test Manual'*, Windsor: NFER-NELSON.

Wechsler, D. (1955), *'Manual for the Wechsler Adult Intelligence Scale'*, New York: The Psychological Corporation.

Wechsler, D. (1958), *'The Measurement and Appraisal of Adult*

Intelligence, 4th ed', Baltimore: Williams & Wilkins.

Wechsler, D. (1981), *'Manual for the Wechsler Adult Intelligence Scale – Revised'*, New York: The Psychological Corporation.

Wechsler, D. (1988), *'Wechsler Memory Scale – Revised'*, San Antonio: Harcourt, Brace, Jovanovich.

Welsh Office (1983), *'All Wales Strategy for the Development of Services for Mentally Handicapped People'*, Cardiff: HMSO.

WHO – World Health Organisation (1991), *'The Neurological Adaptation of the International Classification of Diseases (ICD-10NA)'*, Geneva: World Health Organisation.

Wilson, B.A., Cockburn, J. & Baddeley, A. (1985), *'Rivermead Behavioural Memory Test Manual'*, Bury St Edmunds: Thames Valley Test Co.

Wilson, B.A., Cockburn, J. & Baddeley, A. (1991), *'Rivermead Behavioural Memory Test Manual, 2nd ed'*, Bury St Edmunds: Thames Valley Test Co.

Winblad, B., Cummings, J., Andreasen, N., Grossberg, G., Onofrj, M., Sadowsky, C., Zechner, S., Nagel, J. & Lane, R. (2007), 'A six-month double-blind, randomized, placebo-controlled study of a transdermal patch in Alzheimer's disease - rivastigmine patch versus

capsule', *International Journal of Geriatric Psychiatry, 22;* 5: 456-467.

Wolfensberger, W. & Kugel, R. (1969), *'Changing Patterns in Residential Services for the Mentally Retarded'*, Washington D.C.: President's Committee on Mental Retardation.

Young, E.C. & Kramer, B.M. (1991), 'Characteristics of age-related language decline in adults with Down's syndrome', *Mental Retardation, 29;* 2: 75-79.

Zigman, B.W., Schupf, N., Sersen, E. & Silverman, W. (1996), 'Prevalence of dementia in adults with and without Down syndrome', *American Journal of Mental retardation, 100:* 403-412.

Zigmond, A.S. & Snaith, R.P. (1983), 'The Hospital Anxiety and Depression Scale', *Acta Psychiatrica Scandinavica, 67:* 361-370.

Zimmerman, I.L. & Woo-Sam, J.M. (1973), *'Clinical Interpretation of the Wechsler Adult Intelligence Scale'*, New York: Grune & Stratton.

PART I: Empirical Studies (Published Papers)

Chapter 2

A NEUROPSYCHOLOGICAL TEST BATTERY FOR IDENTIFYING DEMENTIA IN PEOPLE WITH DOWN'S SYNDROME

S.B.N. Thompson

The British Journal of Developmental Disabilities (1994),

XL; 2 (79): 135-142.

ABSTRACT

A test battery comprising 6 assessment tools to assess and monitor cognitive decline in elderly dementing clients with learning disabilities is presented. Findings from the study involving 8 clients with Down's syndrome is detailed together with a discussion of the problems of testing elderly clients with learning disabilities. The test battery presented,

attempts to provide the clinician with a convenient and relatively simple procedure for establishing and monitoring decline in clients with learning disabilities. It is not intended as a definitive measure of Alzheimer's disease or multi-infarct dementia but does provide a standardised format of monitoring decline in a range of cognitive functions over time. Generally, such tools are of considerable assistance and importance, especially when the demands of assessing elderly dementing clients with learning disabilities seem to be on the increase.

INTRODUCTION

Perhaps a reflection of the growing number of elderly people with learning disabilities has been the increasing number of referrals to both institution-based departments of clinical psychology and those that provide community services (Thompson, 1993). Often these referrals have been requests for assessing clients' cognitive abilities. Typically they have been for assessing clients' memory function and orientation for time and place in order to discern whether clients present with some of the signs associated with dementia. Identifying these signs early on

clearly has many advantages including the planning and provision of specialist care for these people.

However, it is often difficult to discriminate between pre- or perinatal brain damage (eg meningitis; anoxia) in association with normal ageing (see Gath, 1986) and those considered to be the result of a dementing process, such as multi-infarcts or Alzheimer's disease (eg Haxby, 1989). The situation is more complicated in people with a learning disability when there can be other confounding variables such as the long-term effects of institutional living, communication and comprehension difficulties, and the lack of a premorbid intelligence quotient since intellectual deficits may have originated from birth. There is also the well-documented proposed link between Down's syndrome and Alzheimer's disease (eg Miniszek, 1983; Kolata, 1985; Oliver & Holland, 1986; Delabar, *et al*., 1987; St Clair, 1987; St George-Hyslop, *et al.*, 1987; Prosser, 1989; Thompson, 1993).

Identifying the clinical signs of dementia usually involves reference to the internationally recognised guidelines of the Diagnostic and Statistical Manual of Mental Disorders – DSM – III – R (APA, 1987). The salient points of the full-length definition (all of which do not necessarily have to be present for the diagnosis of dementia) are:

1. Impairment of short-term and long-term memory;
2. Impairment of abstract thinking;
3. Impaired judgement;
4. Disturbances of higher cortical function (eg aphasia; apraxia; agnosia; constructional difficulty);
5. Personality change;
6. Specific organic factor;
7. Absence of a non-organic factor as a reason for the symptoms (eg major depression).

This definition of dementia is generally accepted by psychologists and psychiatrists and it is also common to distinguish “presenile” dementia from “senile” both by age of onset and also by type of illness. Lishman (1987) describes two types of dementia: arteriosclerotic (which may also occur as a presenile disease) and parenchymatous senile dementia. The latter, which refers to a dementing process in the “parenchyma” or “functional part” of the brain, is by far the commonest form of dementia and is generally characterised by those deficits found in Alzheimer’s disase. Brain lesions in atypical Alzheimer-type patient have “miliary”,

"Fischer", or "neuritic plaques" (Kolata, 1985); neurofibrillary tangles; and degeneration of the ends of nerve cells.

The paradox between unequivocal neuropathological findings and limited clinical evidence of dementia, particularly in Down's syndrome, has been partly resolved by the use of specific neuropsychological assessments to detect age-related deficits (eg Dalton, *et al.*, 1974; Wisniewski, *et al.*, 1978; Miniszek, 1983; Wisniewski, *et al.*, 1983), but conclusive evidence to distinguish clinical features of dementia from normal ageing in people with Down's syndrome is still not available.

In an attempt to address some of these complex issues surrounding the identification of dementia in people with learning disabilities, an examination was conducted that considered a range of specific standardised assessments. Following on from this, a neuropsychological test battery was compiled and tested, which comprised four well known psychological assessment tools and two new scales that had been previously tested only in a limited way. In addition, clients' biographical details and medical histories were collected.

NEUROPSYCHOLOGICAL TEST BATTERY

The test battery consisted of the following:

1. Hampshire Social Services Assessment (HSSA);
2. Dementia Questionnaire for Mentally Retarded Persons (DMR);
3. Hospital Anxiety and Depression Scale (HADS);
4. Raven Coloured Progressive Matrices (RCPM);
5. Wechsler Adult Intelligence Scale – Revised (WAIS-R);
6. Middlesex Elderly Assessment of Mental State (MEAMS).

The two new scales that were used in the test battery were the Hampshire Social Services Assessment (HSS, 1989), and the Dementia Questionnaire for Mentally Retarded Persons (DMR), devised and first piloted by Evenhuis, *et al.* (1990). The former of these scales addressed a number of these self-care issues and was designed as a questionnaire to be completed by the carer. Responses to questions were scored and totalled giving an overall ‘band’ that reflected the level of dependence or relative independence of the client. Scores from using this questionnaire could be used for grouping and comparing clients participating in the study according to their abilities/disabilities and dependence/independence. The second new scale, the DMR, was designed to assess the client’s cognitive

and social skills over time and produced sets of scores from totalling the responses to each question that had been completed by the relative or carer of the client. A significant decline in the difference scores from baseline to re-testing reflected a decline in cognitive and/or social functioning. From piloting the questionnaire in the Netherlands, Evenhuis (1992) produced sets of criteria for identifying scores that may indicate dementia in some of the clients in her study.

A third, well known scale, the Hospital Anxiety and Depression Scale or 'HADS' (Zigmond & Snaith, 1983), was used in a modified way to the original intentions of its authors. Since there is little in the way of standardised tests suitable for assessing depression in people with learning disabilities, this questionnaire was used by the carer, in conjunction with the clinical psychologist in an interview situation. Questions were re-phrased in order to pitch questions at a level of understanding appropriate to the individual client. It is appreciated that the original administration of the HADS was as a self-rating tool but it had become apparent in an earlier pilot study that the majority of clients interviewed either could not read or did not have sufficient ability to comprehend the questions in their original form. Therefore, in the absence of more suitable tests, an attempt was made to assess each client's

psychological status, with respect to depression and/or anxiety. This method was largely very satisfactory since it usually confirmed knowledge about the client gained from other sources such as from observations and recordings in the relevant notes or from carers' knowledge at that time. It was considered important to persevere in addressing this issue since difficulties often arise when there is a differential diagnosis of depression and dementia (see Warren, *et al.*, 1989; Yapa & Roy, 1990). Hence, provision for the HADS in the test battery was considered to be important.

The remainder of the test battery comprised well known psychological tests: RCPM (Raven, *et al.*, 1990); WAIS-R (Wechsler, 1981); and the MEAMS (Golding, 1989) which again was not originally designed for people with learning disabilities but has been used in this study.

METHOD

Clients

Five male and 3 female clients with an average age of 51.1 years (range 44 – 63 years) were selected from people living in their own home or in voluntary organisation group homes in the Southampton area. All clients had been diagnosed on referral as having Down's syndrome with an average IQ of 51.2 (range 47 – 63). All clients were asked if they wished to take part in the study with the necessary consent, and approval having been obtained from the University of Southampton and SW Hants Authority Joint Ethics for Research Committee (ref. 54/93: PNN/1/WGP/SJ 07.04.93).

Drugs

Two of the 8 clients received medication during the study; these drugs were reviewed by the visiting general practitioner and, where possible, kept constant throughout the study. One client received phenytoin and the other carbamazepine, both for epilepsy. Neither client had episodes

immediately prior to any of the assessments. Medication was considered to be successfully managing the condition in each case.

Procedure

An explanation of how to complete each rating scale was given to the carer of each consenting client. Carers were asked to complete the HSSA and DMR for their clients based on their knowledge of each client's abilities during the previous two months. The HADS was completed by the clinical psychologist with the assistance of the carer in an interview situation with the client (as explained previously). The remaining tests were administered by the clinical psychologist on two separate occasions, usually beginning with the WAIS-R and followed by the MEAMS and RCPM on the second occasion (not more than 7 days later). In addition, biographical details about each client were collected at this stage of the study from carers' notes, medical notes (of the visiting doctor) and from appropriate carers and/or relatives.

Six months later each client was approached again for permission to re-test them. The WAIS-R was not repeated at this stage because of the possibility of practice effects, nor was the HSSA repeated, unfortunately,

due to chronic short staffing among carers. However, the parallel version B of the MEAMS was administered and the RCPM, DMR and HADS were all administered as before. At 12 months, the DMR and HADS were again repeated to see if any changes had occurred. The following information was obtained from the test battery and analysed using non-parametric statistics (repeated measures) because of the relatively small sample:

1. HSSA – total score and band reflecting the clients' dependence;
2. DMR – sum of cognitive scores and sum of social scores;
3. HADS – total score for anxiety and total score for depression;
4. RCPM – total number of correct responses and percentile;
5. WAIS-R – verbal IQ, performance IQ and full-scale IQ;
6. MEAMS – total score and number of subtests passed;
7. Biographical details of each client tested.

RESULTS

Five out of 8 clients were found to be in band 2 (and one client, in band 1) of the HSSA indicating "support needed for several aspects of care, supervision and independence training (on average 3 or 4 hours in 24)." (HSS, 1989). Two clients fell in band 5, indicating "greater dependence on carers (on average over 7 or 8 hours in 24)", according to the guidelines for the HSSA (HSS, 1989).

All 8 clients scored within the range 1 – 9 on the RCPM placing them below the 5th percentile which is well below that normally expected for the general population. Six out of 8 clients showed score changes of at least 4 points on their sum of social scores in the DMR. These score changes were between each 6-month assessment for 3 clients and between the 6-month and 12-month assessment for the remaining 3 clients. Using the IQ banding criteria for sum of social scores on the DMR (Evenhuis, 1992), 2 clients showed changes in their DMR social scores in excess of 6.3 points, which is the amount proposed for non-demented clients with Down's syndrome. Results of clients' sum of cognitive scores on the DMR indicated decline in 3 clients whose score changes were equal to or above 7 points between both the 6-month and initial assessment, and also

between the 6-month and 12-month assessment. According to the IQ banding for sum of cognitive scores, 2 clients showed changes in their cognitive scores above those of non-demented clients with Down's syndrome (5.8 points is the cut-off). Two clients were in a higher IQ banding to the remainder of clients and no tabulated values have been provided by Evenhuis (1992) for either SOS or SCS score changes in this IQ grouping.

All clients passed an average of 1.4 subtests of the MEAMS at the first assessment, and an average of 2.1 subtests on the second assessment 6 months later. Averages for total scores achieved on the MEAMS also varied: 14.3 (first assessment) and 16.0 (6-month assessment).

Only two clients showed levels of caseness for anxiety during the study and two clients showed caseness depression.

DISCUSSION

All 8 clients in the study achieved scores on the WAIS-R equivalent to an IQ below 70; the average IQ of these clients (51.2) classifies a person, according to Wechsler (1981), as being 'mentally deficient'. This term

may not necessarily be of particular use and indeed stigmatizes people; however, it is perhaps not too surprising that these clients' performances were consistent with those they achieved on the RCPM, placing them at below the 5th percentile. This is also consistent with the finding that all of the clients had fairly high demands on the amount of support time required by carers, as seen by the HSSA scores and bands.

What is surprising is the relative slight increases in some clients' total scores achieved on the MEAMS; although the number of subtests passed (which can be compared with normative tables) remain within those ranges normally expected of dementing persons in the general population. This finding could not be explained by practice effects since the parallel version of the MEAMS had been administered. It is possible that some minor cognitive skills improved with routine care provided by carers. However, it is clear that at least 6 of the 8 clients tested showed signs of declining skills generally, and that scores achieved on the DMR indicated signs of deterioration in cognitive and social skills in most cases. There was also evidence of score changes in the remaining 2 clients, although according to comparative data available from Evenhuis (1992), these changes were not significant. It is always possible, however, that greater

score changes may occur in these clients at a further 6-month re-test, hence showing possible signs of dementia.

Depression is often found in association with dementia, and as with the clients in this study, two clients exhibited signs normally associated with depression. However, one of these clients showed no signs of decline in social skills (DMR); but with the second client the question should be raised of whether or not the presence of depression has confounded findings that indicate decline in social skills or whether in fact a differential diagnosis is necessary to explain findings for this client? It is difficult to discern the sensitivity of the DMR when used with depressed clients generally; indeed Evenhuis (1992) has suggested caution be used when interpreting results obtained in such situations. This leaves us still with the problem generally of discriminating between those clients who are dementing *only* from those that are depressed *only*. This problem has been long debated and presents itself predominantly when faced with assessing clients with limited communication and comprehension skills, such as those with some types of learning disability. Perhaps the only real answer to this problem is long-term behavioural observation of clients.

The DMR presents as a useful tool indicating general areas of clients' skills that have declined; however, there is a need for a definitive

assessment of depression for these clients in order to discern between the effects of depression and those of dementia. Such limitations to the use of the DMR make additional measures necessary; for example, the MEAMS can provide information that allows a subjective "adjustment" to be made to the results of the DMR. What this means is that the DMR may show a particular client to have global deficits, but specific deficits may be pin-pointed by using a direct client measure such as the MEAMS. Afterall, the DMR is a carer-rated tool and cannot measure directly the client's skills such as performance on a specific task.

The RCPM is again a global measure and seeks to tap the client's intellectual abilities providing the scorer with a percentile according to performance. The RCPM tends not to have ceiling or floor effects with most clients who have a learning disability, unlike the WAIS-R (see Jones, 987; Atkinson, 1991), and is therefore a more popular choice of assessment. However, it does not identify a range of abilities like the WAIS-R or provide a profile of skills for a given client.

CONCLUSIONS

In balance, the test battery presented, attempts to provide the clinician with a convenient and relatively simple procedure for establishing and monitoring decline in clients with learning disabilities. It is not intended as a definitive measure of Alzheimer's disease or multi-infarct dementia but does provide a standardised format of monitoring decline in a range of cognitive functions over time. Generally, such tools are of considerable assistance and importance, especially when the demands of assessing elderly dementing clients with learning disabilities seem to be on the increase.

SUMMARY

A test battery comprising 6 assessment tools to assess and monitor cognitive decline in elderly dementing clients with learning disabilities is presented. Findings from the study involving 8 clients with Down's syndrome is detailed together with a discussion of the problems of testing elderly clients with learning disabilities.

REFERENCES

American Psychiatric Association (1987). *'Diagnostic and Statistical Manual of Mental Disorders – Revised (DSM-III-R)'*. Washington DC, American Psychiatric Association, 78-80.

Atkinson, L. (1991). 'Mental retardation and WAIS-R difference scores'. *Journal of Mental Deficiency Research, 35:* 537-542.

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

Delaber, J.M., Goldgaber, D., Laqmour, Y., Nicole, A., Huret, J-L., de Groucy, J., Brown, P., Gajdusek, D.C. & Sinet, P-M. (1987). 'Beta amyloid gene duplication in Alzheimer's disease and karyotypically normal Down's syndrome'. *Science, 235:* 1390-1392.

Evenhuis, H.M. (1992). 'Evaluation of a screening instrument for dementia in ageing mentally retarded persons'. *Journal of Intellectual Disability Research, 36:* 337-347.

Evenhuis, H.M. Kengen, M.M.F. & Eurlings, H.A.L. (1990). *'Dementie vragenlijst zwakzinnigen'*. Zwammerdam, Hooge Burch.

Gath, A. (1986). 'Ageing and mental handicap'. *Developmental and Child Neurology, 28:* 515-524.

Golding, E. (1989). *'The Middlesex Elderly Assessment of Mental State'*. Bury St Edmunds, Thames Valley Test Company.

Hampshire Social Services – HSS (1989). *'Hampshire Social Services Assessment'*. Southampton, Hampshire Social Services.

Haxby, J.V. (1989). 'Neuropsychological evaluation of adults with Down's syndrome: Patterns of selective impairment in non-demented old adults'. *Journal of Mental Deficiency Research, 33:* 193-210.

Jones, R. (1987). 'Applicability of the WAIS-R: Its use with people with mental handicap'. *Mental Handicap, 15:* 155-158.

Kolata, G. (1985). 'Down's syndrome – Alzheimer's linked'. *Science, 230:* 1152-1153.

Lishman, W.D. (987). *'Organic Psychiatry, 2nd ed'*. Oxford, Blackwell.

Miniszek, N.A. (1983). 'Development of Alzheimer's disease in Down's syndrome individuals'. *American Journal of Mental Deficiency, 85:* 377-385.

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

Prosser, G. (1989). 'Down's syndrome, Alzheimer's disease, and reality orientation'. *Mental Handicap, 17; 2:* 50-53.

Raven, J., Raven, J.C. & Court, J.H. (1990). *'Coloured Progressive Matrices'*. Oxford, Oxford Psychologists Press.

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

St George-Hyslop, P.H., Tanzi, R.E., Polinsky, R.J., Haines, J.L., Nee, L., Watkins, P.C., Meyers, R.H. & Feldman, R.G. (1987). 'The genetic defect causing familial Alzheimer's disease maps on chromosome 21'. *Science, 235:* 885-890.

Thompson, S.B.N. (1993). 'Doubting dementia in Down's syndrome'. *The Psychologist, 6; 9:* 397-400.

Warren, A.C., Holroyd, S. & Folstein, M.F. (1989). 'Major depression in Down's syndrome'. *British Journal of Psychiatry, 155:* 202-205.

Wechsler, D. (981). *'Wechsler Adult Intelligence Scale – Revised'*. New York, The Psychological Corporation.

Wisniewski, K.E., Howe, J., Williams, D.G. & Wisniewski, H.M. (1978). 'Precocious ageing and dementia in patients with Down's syndrome'. *Biological Psychiatry, 13;* 5: 619-627.

Wisniewski, K.E., French, J.H., Rosen, L.J., Koziowski, P.B. & Frankiewicz, A. (1983). 'Basal ganglia calcification in Down's syndrome – another manifestation of premature ageing'. *Annals of the New York Academy of Sciences, 396:* 179-189.

Yapa, P. & Roy, A. (1990). 'Depressive illness and mental handicap: Two case reports'. *Mental Handicap, 18: 19-21*.

Zigmond, A.S. & Snaith, R.P. (1983). 'The Hospital Anxiety and Depression Scale'Acta Psychiatrica Scandinavica, 67: 361-370.

Chapter 3

EXAMINING DEMENTIA IN DOWN'S SYNDROME (DS): DECLINE IN SOCIAL ABILITIES IN DS COMPARED WITH OTHER LEARNING DISABILITIES

S.B.N. Thompson

Clinical Gerontologist (1999), 20; 3: 23-44.

ABSTRACT

Twenty clients with Down's syndrome (DS) and 21 clients with non-Down's syndrome (NDS) learning disabilities were assessed using specially selected neuropsychological assessment tools at two points separated by twelve months. Evidence was found to support hypothesis 1 which suggested that people with DS show a greater decline in social abilities with age, compared with other groups of people with learning

disabilities. Statistically, score changes reflecting the social abilities of the DS clients were found to be significantly greater ($p < .002$) than those of the NDS clients. Findings were explained in terms of poor language abilities in the DS people generally, and the link between declining social abilities and dementia.

INTRODUCTION

Increasing longevity, especially of people with a learning disability (Eyman, *et al.*, 1987; Eyman, *et al.*, 1991), has brought with it a seemingly ever-increasing demand on health and social services. Supportive consultation with staff and clients is important and has also increased the demands on all services as the size of the older population has grown.

Identifying signs of declining memory and general cognitive functioning early on clearly has many advantages (Huppert & Tym, 1986), including the planning and provision of specialist care. Researchers and clinicians have been interested in the effects of ageing on the normal population for some considerable time (Holden,

1989) and have compared common impairments such as short-term memory (McDade & Adler, 1980); age-related memory decline (Young & Kramer, 1991); and psychophysiological differences such as auditory event-related potentials (Muir, *et al.*, 1988). The difficulties of a differential diagnosis between depression and dementia have also been examined (Warren, *et al.*, 1989) but the stumbling block of researchers has often been the transferability of measures to different client groups (Rosen, *et al.*, 1984). Often standardised assessments are too difficult or are culturally-dependent; testing some clients results in floor or ceiling effects; and other tests are simply too demanding of a person's attention or concentration.

Normal Ageing

Normal ageing can be distinguished in terms of biological, social and psychological, but there is often a great overlap and interaction between them. For example, a physical change such as arthritis can limit mobility, which in turn can reduce involvement in social activities or other previous sources of enjoyment (Alcott, 1993). The influence of one aspect of ageing on another should also be remembered; this is important when

considering and comparing past and present cognitive functions within the same person.

Defining "normal" is a difficult task and it is surprising how "normal" and "abnormal" activities and attitudes often overlap. The blurring of boundaries occurs between different cultures, different environments or even between individuals. A misconception is to consider normality as distinct and opposite to abnormality when in fact "normality" refers to the "range around the middle of a dimension (eg height) with two extremes at opposite ends (very tall and very short), rather than one extreme" (Alcott, 1993). Different people have their own opinion about normality and hence expectancies in ageing are perceived differently between individuals. With the advance of medicines and technology, people are generally living longer and so more people are exposed to older people and are witness to the variations in ageing of relatives and friends. In turn, people's understanding of normal ageing is being constantly revised and so too are their expectancies of themselves and others.

Normal ageing brings with it changes, not just to an individual's appearances, however subtle, but also certain changes to the higher mental functions or "cognitive" functions (Allen, *et al.*, 1997). Memory can also be affected (Small, *et al.*, 1995), sometimes because the

individual has failed to receive information correctly, or sometimes because it can no longer be effectively encoded or stored (Nyberg, *et al.*, 1996). The effect of ageing on memory is very often one of the first of the cognitive functions to be noticed by others and can cause considerable distress to the individual and to relatives, close friends and carers. Deterioration in memory functioning is characteristic of dementia (Mitrushina, *et al.*, 1995) but it can also indicate other dysfunctions which should always be considered in any assessment.

DEFINING DEMENTIA

The definition of dementia generally accepted by clinical psychologists and psychiatrists is that outlined in DSM-III-R (APA, 1994). In summary, it states that for a diagnosis of dementia, there should be demonstrable evidence of impairment in short-term and long-term memory. Impairment in short-term memory (ie inability to learn new information) may be indicated by an inability to remember three objects after five minutes. Long-term memory impairment (ie inability to remember information that was known in the past) may be indicated by an

inability to remember past personal information (eg what happened yesterday; birthplace; occupation) or facts of common knowledge (eg past Prime Ministers; well known dates). The salient points of the full-length definition (all of which do not necessarily have to be present for a diagnosis of dementia) are:

1. Impairment of short-term and long-term memory;
2. Impairment of abstract thinking;
3. Impaired judgement;
4. Disturbances of higher cortical function (eg aphasia; apraxia; agnosia; constructional difficulty);
5. Personality change;
6. Specific organic factor;
7. Absence of a non-organic factor as a reason for the symptoms (eg major depression).

Dementia is commonly misunderstood to be a disease when in fact it may be the result of a number of factors, and in some instances it may be reversible. Stokes and Holden (1993) have described “primary dementia” as an extensive, organic impairment of intellect, memory and personality. It occurs in the absence of clouding of unconsciousness (ie without drowsiness) which is acquired, irreversible and progressive.

Over the years, there have been several different definitions of "dementia" and these have varied often according to the viewpoint of the person proposing the definition; for example, from a neuroanatomist's structural viewpoint of from a neuropsychologist's functional viewpoint. Definitions have changed also with the advent of improved technologies such as Computerised Tomography (CT) scanning and Magnetic Resonance Imaging (MRI). Dementias resulting from a stroke, for example, may be defined generally as "vascular" dementia or "multi-infarct" dementia if it is of a more gradual onset; or specifically according to which blood vessels are involved as in "lacunar stroke".

Neuropsychology and Clinical Signs of Alzheimer's Disease

Alzheimer's disease is the single most common form of dementia, accounting for more than 50 per cent of cases of dementia in those over the age of 65 (Katzman, 1976). Initially, the neuropathology of Alzheimer's disease was thought to be arteriosclerotic; however, this was revised after researchers (eg Tomlinson, *et al.*, 1970; Lishman, 1987) consistently reported arteriosclerosis in people with a diagnosis of presenile dementia.

More recently, researchers have described dementias in terms of specific neuropathology (eg Newcombe, 1993 on frontal lobe dementia). Several different findings now enable Alzheimer's disease to be distinguished according to discrete brain abnormalities; for example, Mulder, *et al.* (1998) discovered altered lipid homeostasis in the brain of Alzheimer patients that is not related to the presence of apolipoprotein E4, which affects plasma lipid metabolism. Deb (1997) and Emerson, *et al.* (1995) have confirmed using magnetic resonance imaging that people with Down's syndrome show signs associated with Alzheimer's disease, ie dilation of ventricles, increased peripheral atrophy and increased deep white matter lesions. However, those with Down's syndrome and clinical dementia, show atrophic changes similar to those seen in the general population and non-specific basal ganglia changes (Deb, 1997).

Others have re-defined the stages of Alzheimer's disease (eg Grober, Dickson & Slininski, 1999; for example, Braak & Braak (1991; 1996a,b) discuss the specific pattern of the disease-related lesions. Six stages are distinguished according to location of the tangle-bearing neurons and severity of changes: Stages I; transentorhinal Stages II: clinically silent cases; limbic Stages III-IV: incipient Alzheimer's disease; neocortical Stages V-VI: fully developed Alzheimer's disease. Braak, *et al.* (2000)

explains that in Alzheimer's disease the illness remains subclinical for years with clinical symptoms only being observed late in the course, and their appearance is usually concurrent with the encroachment of the destructive process upon neocortical association areas.

DEFINING LEARNING DISABILITY

Two per cent of the UK population (over one million people) have learning disabilities, the majority of them mild. "Learning disability" is a very broad term and has been used to describe people with an intelligence quotient (IQ) below 70. Wechsler (1981) has classified the abilities of groups of people according to IQ (Table 1). People with learning disabilities commonly may have a range of difficulties which might include approaches to problem-solving, coordination difficulties, problems with speech or comprehension, cognitive delay, or slowness or inability to perform daily routines, such as hygiene or feeding (Thompson, 1993a). The range or number of difficulties an individual may have can be very large or, equally, very small. Increasingly, therefore, it has been useful to state a person's abilities rather than emphasising their negative disabilities. With the promotion of

TABLE 1: Intelligence classifications (adapted from Wechsler, 1981)

Intelligence Quotient (IQ)	Classification
130 and above	Very Superior
120 - 129	Superior
110 - 119	High Average
90 - 109	Average
80 - 89	Low Average
70 - 79	Borderline
69 and below	Mentally Retarded

community living, definitions of learning disability have come to include the extent of a person's ability to live alone or his or her "independence".

A useful, working definition (Thompson, 1993b) states:

> A person with a learning disability is someone who is, to a varying degree, dependent on others for their living needs because of a cognitive impairment resulting from hereditary abnormalities or directly following (or during) birth. They may (or may not) also have associated physical/sensory/behavioural/ medical disabilities. (p. 195)

Definition of Down's Syndrome

"Down's syndrome" (previously known as "mongolism") in one of the most frequently observed forms of learning disability (Moody & Moody, 1992). The former is used more often clinically and was first described by Langdon Down as a separate entity in 1866 (Down, 1866) and independently in the same year by Seguin (Seguin, 1866). Seguin referred to the disorder as "furfuraceous cretinism", emphasising an

assumed relationship to cretinism, while Down, struck by some aspects of the physiognomy of the patients which were superficially similar to those of people in outer Mongolia, called it Mongolian idiocy. Thankfully today people with such disorders are more commonly referred to by their first names, thus recognising the fact that there is a person behind such stigmatizing labels.

A characteristic of Down's syndrome is the presence of an extra gene on chromosome 21 (i.e., "trisomy 21") (Shermann, *et al.*, 1991). Often the person has developmental delays (Maclean, *et al.*, 1991), sometimes a slightly larger head circumference (Palmer, *et al.*, 1992), and language difficulties, especially with ageing (Young & Kramer, 1991). Down's syndrome has been the focus of much research and controversy (Barr, 1990); because of increased longevity, individuals with Down' syndrome are living long enough to be at risk for a host of age-related diseases, for example, pre-senile dementia (Wisniewski, *et al.*, 1983).

NEUROPSYCHOLOGY OF DEMENTIA WITH DOWN'S SYNDROME

It should be noted that, just as with the general population, people with learning difficulties can develop any of the different types of dementias, ie vascular dementia, arteriosclerotic or parenchymatous (such as Alzheimer's disease). As with the wider "normal" population, these different diseases have different courses, for example, step-wise versus insidious decline when comparing multi-infarct dementia with Alzheimer's disease. There is no evidence to suggest that the neuropsychology of dementia is any different in the learning disability population excepting the difference in level of performances obtained on neuropsychological tests. These performances will depend upon the level of general abilities possessed by the client being tested.

A number of difficulties arise when attempting to assess people with a learning disability, whether or not they possess the signs associated with dementia. For example, many of these clients have limited or poorly developed language; poor comprehension; apraxia or agnosia; or suffer from depressive illness (Cooper & Collacott, 1993); or other psychiatric disabilities (Moss, *et al.*, 1991). They may have specific physical

disabilities, such as incontinence, abnormal reflexes, or behavioural problems (eg inappropriate behaviour, stereotyped movements, or pronounced anxiety) that make conventional psychological testing awkward or even impossible. These clients may simply manifest the processes of normal ageing (Hogg, *et al.*, 1988).

Combining cognitive tests with other measures of dementia pathology (eg computerised axial tomography scans - Schapiro et al., 1987) may reveal more distinctive early clinical indicators of deterioration. Indeed, St Clair and Blackwood's (1985) finding that evoked potential latency in those with Down's syndrome increased significantly earlier in life than normal controls, may provide this early indication when correlated with cognitive test results. In the absence of such technology, conventional psychological techniques must be used, but careful selection and possible modification of test batteries becomes necessary when assessing people with learning disabilities.

In 1876, Fraser and Mitchell first noted an association between Down's syndrome and Alzheimer's disease, but it was not until 1929 when Struwe described the characteristic senile plaques of Alzheimer's disease in the brains of individuals with Down's syndrome (Oliver & Holland, 1986). Verhaart and Jelgersma (1952) described clinical

deterioration associated with Alzheimer-like changes at post mortem in a number of people with Down's syndrome; subsequent research focused on establishing similarities between the neuropathological changes in the brains of elderly Down's syndrome individuals and the senile plaques, neurofibrillary tangles and granulovascular degeneration characteristic of Alzheimer's disease. By the 1960s the link between the two disorders was clearly established; and it was agreed that all people with Down's syndrome over the age of 35 have the neuropathological features of Alzheimer's disease (Heston, 1977).

However, there is still debate over whether or not people with Down's syndrome also present with typical clinical features of Alzheimer's disease (Roper & Williams, 1980), since many such individuals maintain good physical an mental health into the fourth an fifth decades of life (St Clair & Blackwood, 1985). Interest in the relationship between Alzheimer's disease and Down's syndrome has been further stimulated by discoveries localising two distinct Alzheimer's disease markers to chromosome 21. First, the gene encoding the precursor protein which, in some processed form, gives rise to the amyloid deposits in Alzheimer's disease is found on chromosome 21 (Kang, *et al.*, 1987). Second, a marker has been identified on chromosome 21 which is linked to within

15 centrimorgans (a distance spanning 300 600 genes or 15 million base pairs) of a site associated with an automosal dominant form of Alzheimer's disease (St George-Hyslop, *et al.*, 1987). Arguments in favour of some kind of connection have been advanced for some time (Prosser, 1989), but evidence from some studies also puts doubt on such a definitive connection; for example, it is suggested that the amyloidogenic gene or chromosome 21 is not identical to the Alzheimer's gene despite the widespread presence of amyloid material in the senile plaques and neurofibrillary tangles (Curran & Wattis, 1989). This might imply the picture reflected in Down's syndrome may be of a different brain process to that evidenced in sufferers of Alzheimer's disease.

Although there is little definitive evidence of a family history of Alzheimer's disease in sufferers, an assumed inheritance of Alzheimer's disease within families has been reported in a few studies (Whalley, *et al.*, 1982; Breitner & Fostein, 1984). However, these studies are difficult to compare and often have poor follow-up of subjects or use different criteria for the diagnosis of dementia (McKusick, 1983). Studies of twins have been used to try and disentangle the roles of environment and genetics. In one such study, the incidence of dementia for monozygotic twins was found to be less than for dizogotic twins (Deary & Whalley,

1988). Thus, although genetic factors are important, environmental factors seem to play an essential role.

In some neuropathological studies of Down's syndrome it has been suggested that the present diagnostic clinical signs of Alzheimer's disease are easily applicable (Reid, *et al.*, 1978). While others argue that the signs of deterioration associated with ageing in Down's syndrome need to be clarified before the diagnosis of Alzheimer's disease can be generally accepted (Sylvester, 1984). The major difficulty in applying criteria is the underlying learning disability. As Levinson and colleagues (1955) have commented, there is great variation in the developmental theories and in the intelligence quotients (IQs) of people with Down's syndrome (and other learning disabilities). Thirty per cent of subjects studied had an IQ of less than 20,65 per cent between 20 and 50, and five per cent between 50 and 65 (Breg, 1977). These hold great restraints on the application and interpretation of conventional test procedures (Atkinson, 1991): failure or poor performance might indicate dementia or an underlying disability. Clearly, these restrictions would not be necessary if the "intelligence" of this client population was evenly distributed throughout age groups.

When considering these issues, other questions are raised. For instance, are the associated changes in behaviour present but undetected

in people with Down's syndrome due to poor institutional environments? To complicate this further, it also seems likely that there is an increased prevalence of epilepsy with age in sufferers of Alzheimer's disease (Tangye, 1979). Making the distinction between the effects of long-term epilepsy and types of brain damage on cerebral function can often be a difficult process (Thompson, *et al.*, 1992).

Although a high proportion of individuals with Down's syndrome develop the neuropsychological changes of Alzheimer's disease, only a proportion develop the definite signs of deterioration and have the clinical features characteristic of the latter stages of Alzheimer's disease. It is also often difficult to discriminate between pre- or perinatal brain damage (eg meningitis; anoxia) in association with normal ageing (Gath, 1986) and those considered to be the result of a dementing process (Haxby, 1989). The situation is more complicated in people with a learning disability when there can be other confounding variables such as the long-term effects of institutional living, communication and comprehension difficulties, and the lack of a premorbid intelligence quotient since intellectual deficits may have originated from birth.

The paradox between unequivocal neuropathological findings and limited clinical evidence of dementia, particularly in Down's syndrome,

has been partly resolved by the use of specific neuropsychological assessments to detect age-related deficits (Miniszek, 1983) but conclusive evidence to distinguish clinical features of dementia from normal ageing in people with Down's syndrome is still not available.

ASSESSING MEMORY AND COGNITIVE FUNCTIONING

Applicability of test material that is already available and standardised, is an important consideration but the transferability of norms across different client groups, for example using the WAIS-R (Wechsler, 1981), has also been debated.

Tests, such as the Middlesex Elderly Assessment of Mental State (Golding, 1989), are a very useful screening tool for dementia and can help indicate the need for further testing in specific areas of deficits, such as memory for faces, recognition of everyday objects or arithmetic ability.

Lishman (1987) warns that unless a full and comprehensive evaluation is made of a suspected dementing patient, the label of a primary dementing illness, for example, carries a hopeless prognosis. Care must be taken not only in carrying out tests but also in the interpretation of

results. Hence, a specific and detailed knowledge base is required of a clinician to interpret results even if the actual tests have been carried out by non-specialised generic workers.

METHOD

Rationale and Hypothesis

There is substantial evidence to show that cognitive abilities may decline with the onset and progress of dementia. A link has been found between people who have the neuropathology of Down's syndrome and their subsequent development of clinical signs, associated with Alzheimer's-type dementia. Less is known about the effects of dementia on non-Down's syndrome persons with learning disabilities and whether there is any difference when compared with Down's syndrome individuals.

Comparing cohorts from other studies that have examined dementia in people with learning disabilities (Crayton & Oliver, 1993), a five-to six-month period between test and retest is quite typical and generally

acceptable. In fact, a twelve-month follow-up was chosen in order to maximize any possible changes and effects, such as cognitive decline.

Hypothesis 1

People with Down's syndrome show a greater decline in social abilities with age, compared with other groups of people with learning disabilities.

Materials

Wechsler Adult Intelligence Scale-Revised (WAIS-R); Raven Coloured Progressive Matrices (RCPM); Hampshire Social Services Assessment (HSSA); Dementia Questionnaire for Mentally Retarded Persons (DMR); Hospital Anxiety and Depression Scale (HADS); information sheets for participants; sheets for the collection of clients' biographical details, and consent forms.

Sample

(i) Group 1: "Down's Syndrome" (DS)

Eleven male and 9 female clients (DS1-DS20) with an average age of 47.25 years (sd = 1.33) and range of 35-63 years (Table 2) were randomly selected from the consultant psychiatrist's lists of people living in the South of the UK. All clients agreed to participate in the study.

Intellectual Functioning

All clients were diagnosed by the consultant psychiatrist as having Down's syndrome and were found to have an average full-scale IQ on the WAIS-R of 51.90 (sd = 1.21) and age range of 47-63 years (Table 2). Scores of clients' performances on the RCPM (Raven, *et al.*, 1995) placed the majority of clients in the "less than the 5th percentile" band, indicating low intellectual ability.

Level of Independence

Norms for the HSSA scores are allocated 'bands'. For the majority of clients with HSSA scores of 130, these fell into band 1, indicating "very minimal support needed (i.e. up to 1 hour in 24, often less)" (HSS, 1989; p 7). Five clients fell into band 2, and two clients fell into band 5; the latter indicating "support needed in all major areas (on average over 7/8 hours in 24). Sleeping-in staff or waking staff needed." Mean band: 1.65; sd: 3.03; range: 1-5 (Table 2).

(ii) Group 2: "Non-Down's Syndrome" (NDS)

Ten male and 11 female clients (NDS1-NDS21) with a range of age of 56.43 (sd = 1.21) years and age range of 45-68 years (Table 3) were randomly selected from the consultant psychiatrist's lists of people living in the South of the UK.

All clients were diagnosed by the consultant psychiatrist as having a learning disability other than Down's syndrome. Clients fell into the following broad categories: prenatal abnormality/illness (eg rubella;

TABLE 2: Group 1: Summary of HSSA and WAIS-R results

CLIENTS	HSSA		AGE	WAIS-R (IQ)		
	Score	Band		Verb.	Perf.	F-Scale
MEAN	37.83	1.65	47.25	54.75	59.00	51.90
SD	4.86	3.03	1.33	1.32	1.12	1.21

Key: Verb. = Verbal IQ
Perf. = Performance IQ
F-Scale = Fullscale IQ
SD = Standard Deviation

TABLE 3: Group 2: Summary of HSSA and WAIS-R results

CLIENTS	HSSA		AGE	WAIS-R (IQ)		
	Score	Band		Verb.	Perf.	F-Scale
MEAN	55.19	2.29	56.43	61.90	63.43	59.19
SD	2.29	1.75	1.21	1.18	1.14	1.17

Key: Verb. = Verbal IQ
SD = Standard Deviation
Perf. = Performance IQ
F = Full-scale IQ

meningitis); perinatal trauma (eg anoxia; cerebral palsy); or metabolic (eg phenylketonuria). None of the clients had a genetically acquired learning disability. All agreed to participate in the study.

Intellectual Functioning

Clients were found to have an average full-scale IQ on the WAIS-R of 59.19 (sd = 1.17) and range of 51-69 years (Table 3). Scores of clients' performances on the RCPM placed the majority of clients in the "less than the 5th percentile" band, indicating low intellectual ability; five clients were placed in the higher 25th percentile ability band.

Level of Independence

HSSA scores of then clients fell into band 3, indicating "support for several aspects of care, supervision and independence training, on average 3 or 4 hours in 24". Six clients fell into band 1, four clients fell into band 2, and one client fell into band 4; the latter indication "support for several aspects of care". Mean band; 2.29; sd: 1.75; range: 1-4 (Table 3).

Controls

Each client was his/her own control. Individual's scores obtained at the first assessment were compared with their own scores at each subsequent assessment. In this way, it was possible to identify those clients who showed probable decline in cognitive and social abilities, as reflected in their change in scores over time.

Consent

Permission and approval for this study was granted by the local ethics committees. Consent was obtained from those people in charge of the caring establishments approached as well as the clients themselves.

Procedure

An explanation of how to complete each rating scale was given to the carer of each consenting client. Carers were asked to complete the HSSA and DMR for each of their clients' abilities during the previous two months. The remaining tests were administered by the clinical

psychologist. At 12 months, each client was approached again for permission to re-test them. The WAIS-R was not repeated at this stage in order to avoid practice effects, nor was the HSSA because of a chronic staff shortage among carers. However, the RCPM, DMR and HADS were administered as before.

The following summarizes information obtained from each assessment:

First Assessment

1. HSSA - total score and band reflecting the client's dependence;
2. DMR - sum of cognitive scores and sum of social scores;
3. HADS - total score for anxiety and total score for depression;
4. RCPM - total number of correct responses and percentile;
5. WAIS-R - verbal IQ, performance IQ and full-scale IQ;
6. Biographical details of each client tested.

Second Assessment

1. DMR - sum of cognitive scores and sum of social scores;

2. HADS - total score for anxiety and total score for depression;
3. RCPM - total number of correct responses and percentile;

RESULTS

Scores obtained using the RCPM reflected performances below the 5th percentile in all but one case for each client group (DS17 & NDS9), which were both at the 10th percentile. Test scores obtained for each group of clients were compared. No statistically significant differences were found between the measure; Age, WAIS-R, Full-scale IQ and HSSA band thus confirming the similarity of ability profiles of the two groups.

Using Analysis of Variance, two measures were found to be significantly different between the client groups, the "sum of social 'difference' scores" of the DMR ($p < .002$; $df = 9$; $F = 8.222$), and the "sum of social scores" at assessment 2 ($p < .05$; $df = 13$; $F = 4.134$). There was a greater difference in scores for the Down's syndrome group as compared with the non-Down's syndrome group. After partialling out HADS Anxiety and HADS Depression scores (Table 4), a significant difference remained for the "sum of social 'difference' scores" ($p < .02$, HADS Anxiety partialled out; $p < .002$, HADS Depression partialled out).

TABLE 4: Comparison of test measures between Group 1 and Group 2 at assessments 1 and 2 controlling for 'anxiety' and 'depression'

Variable	Anxiety Out			Depression Out		
	df	F	p	df	F	p
SOS diff.	9	7.526	<.02	9	14.419	<.003
SCS diff.	2	0.971	.404 (ns)	2	0.615	.556 (ns)
SOS (ass.1)	15	1.347	.507 (ns)	15	0.499	.831 (ns)
SCS (ass.1)	5	1.661	.218 (ns)	5	1.153	.386 (ns)
SOS (ass.2)	13	3.234	.133 (ns)	13	3.456	.120 (ns)
SCS (ass.2)	12	0.604	.780 (ns)	12	1.129	.480 (ns)

Key: SOS (SCS) = Sum of social (cognitive) scores of DMR
diff. = Difference between scores at assessment 1 and 2
ass.1 = Assessment 1 (2)
ns = Not significant

No significant interaction effects were found. These findings support the hypothesis which suggests there is a difference between the two client groups in their social abilities.

DISCUSSION

Although the majority of HADS scores for clients fell within the "normal" range (ie below 8 points), the effect of controlling for these scores was evidenced when they were partialled out in data analyses; $p < .02$ (after HADS Anxiety was partialled out), and $p < .003$ (after HADS Depression was partialled out). Therefore, the level of statistical significance of this finding was reduced following controlling for the HADS scores.

It is perhaps not surprising to find that anxiety and depression are related to the level of social abilities. This finding tends to support what is already known about the influence of depression and anxiety levels on clients' abilities (Agbayewa, *et al.*, 1991) and it is suspected that although the HADS scores for the majority of clients in the study were not in the "borderline" or "caseness" range, some level of anxiety and/or depression

does seem to affect a person's capabilities and performance in the area of social skills.

In addition, the Down's syndrome clients showed a greater score difference between assessments 1 and 2 in the "sum of social scores" than the non-Down's syndrome clients. This supports hypothesis 1 which proposed that people with Down's syndrome show a greater decline in social abilities with age, compared with other groups of people with learning difficulties.

There was also a greater variance in the scores of the Down's syndrome clients. It is known that people with Down's syndrome have developmental delays (Maclean, *et al.*, 1991) and very often also have expressive (Young & Kramer, 1991) and receptive (Hartley, 1982) language difficulties. Therefore, it is proposed that the differences in scores between people with Down's syndrome and other learning disabilities may be due partly to the known differences in understanding language and in the ability for expressive language. Since language is important in the development of social abilities, this impediment may well account for the difference in social abilities seen between the two client groups.

The finding that the Down's syndrome clients showed a greater score difference between assessments 1 and 2 on this measure is interesting in light of reports that suggest Down's syndrome people have neuropsychological profiles similar to those who have been diagnosed with Alzheimer's disease (Roper & Williams, 1980). Such clients have also shown signs of declining social skills. Other reports support the view that changes in a person's personality, such as sociability and social skill evidenced when they were partialled out in data analyses; $p < .02$ (after HADS Anxiety was partialled out), and $p < .003$ (after HADS Depression was partialled out). Therefore, the level of statistical significance of this finding was reduced following controlling for the HADS scores.

It is perhaps not surprising to find that anxiety and depression are related to the level of social abilities. This finding tends to support what is already known about the influence of depression and anxiety levels on clients' abilities (Agbayewa, *et al.*, 1991) and it is suspected that although the HADS scores for the majority of clients in the study were not in the "borderline" or "caseness" range, some level of anxiety and/or depression does seem to affect a person's capabilities and performance in the area of social skills.

In addition, the Down's syndrome clients showed a greater score difference between assessments 1 and 2 in the "sum of social scores" than the non-Down's syndrome clients. This supports hypothesis 1 which proposed that people with Down's syndrome show a greater decline in social abilities with age, compared with other groups of people with learning difficulties.

There was also a greater variance in the scores of the Down's syndrome clients. It is known that people with Down's syndrome have developmental delays (Maclean, *et al.*, 1991) and very often also have expressive (Young & Kramer, 1991) and receptive (Hartley, 1982) language difficulties. Therefore, it is proposed that the differences in scores between people with Down's syndrome and other learning disabilities may be due partly to the known differences in understanding language and in the ability for expressive language. Since language is important in the development of social abilities, this impediment may well account for the difference in social abilities seen between the two client groups.

The finding that the Down's syndrome clients showed a greater score difference between assessments 1 and 2 on this measure is interesting in light of reports that suggest Down's syndrome people have

neuropsychological profiles similar to those who have been diagnosed with Alzheimer's disease (Roper & Williams, 1980). Such clients have also shown signs of declining social skills. Other reports support the view that changes in a person's personality, such as sociability and social skill performance, is affected in people with Down's syndrome who have been diagnosed with Alzheimer's disease (Reid, *et al.*, 1978).

In comparison with Evenhuis' (1992a,b) samples in her evaluation of the DMR, for both client groups in this study, the means of DMR "sum of social scores" were higher; therefore, they had a better range of social skills to begin with (as indicated by their DMR scores) and were generally more able. Yet, it is interesting to note that 15 clients from the two groups showed a significant decline in these scores between assessments 1 and 2.

Eighteen clients in the Down's syndrome group and 19 clients in the non-Down's syndrome group showed changes in their DMR "sum of social scores" between assessments 1 and 2. Of the Down's syndrome clients, nine fulfilled Evenhuis' (1992b) criterion for the diagnosis of a dementing process, ie they had a 4-point score change between assessments. Of the non-Down's syndrome clients, six showed a 4-point score change. Evenhuis (1992a) includes a further set of criteria related

to IQ level. For the "high-moderate level of retardation (IQ 45-55)", the change in "sum of social scores" is given as greater than or equal to 15. Three clients from the Down's syndrome clients (DS8, DS13 and DS14) fell into this category. The mean IQ of the non-Down's syndrome clients was slightly higher than for the Down's syndrome group; therefore, Evenhuis' (1992a) "mild-moderate level of retardation (IQ = 55-70)" applies where the change in "sum of social scores" is given as greater than or equal to 10. In the non-Down's syndrome group, one client (NDS16) fell into this category. Overall, the Down's syndrome group had more clients who had greater changes in their "sum of social scores" between assessments 1 and 2.

CONCLUSIONS

A statistically significant difference ($p < .002$) was found in the "sum of social scores" on the Dementia Questionnaire for Mentally Retarded persons - DMR (ie "social abilities") between Down's syndrome and non-Down's syndrome clients between assessments 1 and 2. This decline was greater in the Down's syndrome clients. Hence, support was found for our

hypothesis, which proposed that the Down's syndrome clients showed a greater decline in social abilities with age, compared with other groups of people with learning disabilities.

Several explanations were proposed for the differences found between the two client groups; namely, the well-documented difficulties with language (comprehension and expression) in people with Down's syndrome may affect their social abilities. Also, in comparison with Evenhuis' (1992a,b) samples, decline in social abilities has been found to be associated with a dementing process particularly in people with Down's syndrome.

REFERENCES

Agbayewa, M.D., Weir, J., Tuokko, H. & Beattie, L. (1991), 'Depression in dementia: Its impact on functional ability', *Dementia, 2:* 212-217.

Alcott, D. (1993), 'Normal Aging' in Stokes, G. & Goudie, F. (eds), *Working with Dementia,* Oxford: Winslow Press, 9-15.

Allen, P.A., Smith, A.F., Jerge, K.A. & vires-Collins, H. (1997), 'Age differences in mental multiplication: evidence for peripheral but not central decrements', *Journal of Gerontology , 52;* 2: P81-P90.

APA-American Psychiatric Association (1994), '*Diagnostic and Statistical Manual of Mental Disorders (DSM-IV). 4th ed'*, Washington DC: American Psychiatric Association, 139-143.

Atkinson, L. (1991), 'Mental retardation and WAIS-R difference scores', Journal of *Mental Deficiency Research, 35;* 537-542.

Barr, O. (1990), 'Down's syndrome and Alzheimer's disease - what is the link?' *Professional Nurse, June;* 465-468.

Braak, H. & Braak, E. (1991), 'Neuropathological staging of Alzheimer-related changes', *Acta Neurophologica, 82:* 239-259.

Braak, H. & Braak, E. (1996a), 'Staging of Alzheimer's disease-related neurofibrillary changes', *Neurobiology of Aging, 16;* 3: 271-282.

Braak, H. & Braak, E. (1996b), 'Mismatch between plaques and tangles in staging Alzheimer pathology: response', *Neurobiology of Aging, 16;* 3: 284.

Braak, H., Griffing, K., Arai, K., Bohl, J., Bratzke, H. & Braak, H. (2000), 'Neuropathology of Alzheimer's disease: What is new since

A Alzheimer?' *European Archives of Psychiatry & Clinical Neuroscience, 249 (supppl 3):* 14-22.

Breg, W.R. (1977), 'A review of recent progress in research', *Pathobiology Annals, 7;* 257-303.

Breitner, J.C.S. & Folstein, M.F. (1984), 'Familial Alzheimer dementia: A prevalent disorder with specific clinical features', *Psychological Medicine, 14;* 63-80.

Cooper, S-A. & Collacott, R.A. (1993), 'Prognosis of depression in Down's syndrome', *Journal of Nervous & Mental Disease, 181;* 204-205.

Crayton, L. & Oliver, C. (1993), 'Assessment of Cognitive Functioning in Persons with Down's Syndrome who develop Alzheimer's Disease' in Berg, J., Karlinsky, H. & Holland, A. (eds), *Down's Syndrome and Alzheimer's Disease,* Oxford University Press.

Curran, S. & Wattis, J.P. (1989), 'Round-up: Searching for the cause of Alzheimer's disease', *Geriatric Medicine, March;* 13-14.

Deary, I.J. & Whalley, L.J. (1988), 'Recent research on the causes of Alzheimer's disease', *British Medical Journal, 297;* 807-809.

Deb, S. (1997), 'Structural neuroimaging in learning disability'. *British Journal of Psychiatry, 171:* 417-419.

Down, J.L. (1866), *'Observations on an ethnic classification of idiots'*, London Hospitals Reports, London: London Hospitals.

Emerson, J.F., Kesslak, J.P. & Chen, P.C. (1995), 'Magnetic resonance imaging of the aging brain in Down's syndrome', *Progress in Clinical & Biological Research, 393:* 123-138.

Evenhuis, H.M. (1992a), *'Provisional Manual of the Dementia Questionnaire for Mentally Retarded Persons (DMR)'*, Zwammerdam: Hooge Burch.

Evenhuis, H.M. (1992b), 'Evaluation of a screening instrument for dementia in ageing mentally retarded persons', *Journal of Intellectual Disability Research, 36:* 337-347.

Eyman, R.K., Call, T.L. & White, J.F. (1991), 'Life expectancy of persons with Down's syndrome', *American Journal of Mental Retardation, 95:* 603-612.

Eyman, R.K., Grossman, H.J., Tarjan, G. & Miller, C.R. (1987), 'Life expectancy and mental retardation. A longitudinal study in a state residential facility', *Monographs of the American Association on Mental Deficiency, 7:* 1-73.

Gath, A. (1986), 'Ageing and mental handicap', *Developmental & Child Neurology, 28:* 515-524.

Golding, E. (1989), *'The Middlesex Elderly Assessment of Mental State'*, Bury St Edmunds: Thames Valley Test Company.

Grober, E., Dickson, D. & Sliwinski, M.J. (1999), 'Memory and mental status correlates of modified Braak staging', *Neurobiology & Aging, 20:* 573.

Hartley, X.Y. (1982), 'Receptive language processing in Down's syndrome children', *Journal of Mental Deficiency Research, 26;* 263-269.

Hartley, X.Y. (1982), 'Receptive language processing in Down's syndrome children', *Journal of Mental Deficiency Research, 26:* 263-269.

Haxby, J.V. (1989), 'Neuropsychological evaluation of adults with Down's syndrome: Patterns of selective impairment in non-demented old adults', *Journal of Mental Deficiency Research, 33;* 193-210.

Heston, L.L. (1977), 'Alzheimer's disease, trisomy 21, and myeloproliferative disorders: Associations suggesting a genetic diathesis', *Science, 196:* 322-323.

Hogg, J., Moss, S. & Cooke, D. (1988), *'Ageing and Mental Handicap'*, London: Chapman & Hall.

Holden, U. (1989), *'Neuropsychology and Ageing'*, London: Chapman &

Hall.

HSS - Hampshire Social Services (1989), *'Hampshire Social Services Assessment'*, Southampton: Hampshire Social Services.

Huppert, F.A. & Tym, E. (1986), 'Clinical and neuropsychological assessment of dementia', *British Medical Bulletin, 42;* 1: 11-18.

Kang, J., Lemaire, H.G., Unterbeck, A., Selbaum, J.M., *et al.,* (1987), 'The precursor of Alzheimer's disease amyloid A4 protein resembles a cell surface receptor', *Nature, 235:* 733-736.

Katzman, R. (1976), 'The prevalence and malignancy of Alzheimer's disease', *Archives of Neurology, 33:* 217-8.

Katzman, R. & Karasu, T.B. (1975), 'Differential diagnosis of dementia' in Fields, W.S. (ed), *Neurological and Sensory Disorders in the Elderly,* Miami: Symposia Specialists, 103-134.

Levinson, A., Friedman, A. & Stamps, F. (1955), 'Variability of mongolism', *Paediatrics, 16:* 43-54.

Lishman, W.A. (1987), *'Organic Psychiatry: the Psychological Consequences of Cerebral Disorder, 2nd ed'*, Oxford: Blackwell.

Maclean, W.E., Ellis, D.N., Galbreath, H.N., Halpern, L.F., *et al.*. (1991), 'Rhythmic motor behaviour of preambulatory motor impaired, Down syndrome and nondisabled children - A comparative analysis',

Journal of Abnormal Child Psychology, 19: 319-330.

McDade, H.L. & Adler, S. (1980), 'Down syndrome and short-term memory impairment: A storage or retrieval deficit?' *American Journal of Mental Deficiency, 84;* 6: 561-567.

McKusick, V.A. (1983), *'Mendelian Inheritance in Man, 6th edn'*, Baltimore: The Johns Hopkins University Press, 30-31.

Miniszek, N.A. (1983), 'Development of Alzheimer's disease in Down's syndrome individuals', *American Journal of Mental Deficiency, 85:* 377-385.

Mitrushina, M., Uchiyama, C. & Satz, P. (1995), 'Heterogeneity of cognitive profiles in normal aging: implications for early manifestations of Alzheimer's disease', *Journal of Clinical and Experimental Neuropsychology, 17;* 3: 374-382.

Moody, P. & Moody, R. (1992), *'Half Left'*, Oslo: Dreyers Forlag A/S.

Moss, S., Goldberg, D. & Patel, P. (1991), *'Psychiatric and physical morbidity in older people with severe mental handicap'*, Report, University of Manchester: Hester Adrian Centre.

Muir, W.J., Squire, I. & Blackwood, D.H.R. (1988), 'Auditory P300 response in the assessment of Alzheimer's disease in Down's syndrome: A 2-year follow-up study', *Journal of Mental Deficiency*

Research, 32: 455-463.

Mulder, M., Ravid, R., Swaab, D.F., de Kloet, E.R., Haasdijk, E.D., Julk, J., van der Boom, J. & Havekes, L.M. (1998), ‘Reduced levels of cholesterol, phospholipids, and fatty acids in cerebrospinal fluid ofAlzheimer disease patients are not related to apolipoprotein E4’, *Alzheimer Disease and Associated Disorders, 12;* 3: 198-203.

Newcombe, F. (1993), ‘Frontal Lobe Disorders’ in Greenwood, R., Barnes, M.P., McMillan, T.M. & Ward, C.D. (eds), *Neurological Rehabilitation,* Churchill Livingstone: Edinburgh.

Nyberg, L., Backman, L., Erngrund, K., Olofsson, U. & Nilsson, L.G. (1996), 'Age differences in episodic memory, semantic memory, and priming: relationships to demographic, intellectual, and biological factors', *Journal of Gerontology, 51;* 4: P234-P40.

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

Palmer, C.G.S., Crank, C., Pueschel, S.M., Wisniewski, K.E., *et al.* (1992), 'Head circumference of children with Down's syndrome (0-36 months)', *American Journal of Medical Genetics, 42:* 61-67.

Prosser, G. (1989), 'Down's syndrome, Alzheimer's disease, and reality orientation', *Mental Handicap, 17;* 2: 50-53.

Raven, J.C., Court, J.H. & Raven, J. (1995), *'Raven Manual: Section 2. Coloured Progressive Matrices 1995 Edition'*, Oxford: Oxford Psychologists Press.

Reid, A.H., Maloney, A.F.J. & Aungle, P.G. (1978), 'Dementia in ageing mentally defectives: A clinical and neuropathological study', *Journal of Mental Deficiency Research, 22:* 233-241.

Roper, A.H. & Williams, R.S. (1980), 'Relationship between plaques, tangles and dementia in Down's syndrome', *Neurology, 30:* 639-644.

Rosen, W.G., Mohs, R.C. & Davis, K.L. (1984), 'A new rating scale for Alzheimer's disease', *American Journal of Psychiatry, 141;* 11: 1357-1364.

Schapiro, M.B., Luxemberg, J.S., Kaye, J.A., Haxby, J.V., *et al.*. (1987), 'Serial quantitative computed tomography analysis of brain morphometry in adult Down's syndrome at different ages', *Annals of Neurology, 22:* 432.

Seguin, E. (1866), *'Idiocy: Its Treatment by the Physiological Method'*, New York: William Wood.

Shermann, S.L., Takesu, N., Freeman, S.B., Grantham, M., *et al.* (1991), 'Association between reduced recombination with nondisjunction', *American Journal of Human Genetics, 49:* 608-620.

Small, G.W., La Rue, A., Komo, S., Kaplan, A. & Mandelkern, M.A. (1995), 'Predictors of cognitive change in middle-aged and older adults with memory loss', *American Journal of Psychiatry, 152;* 12: 1757-1764.

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

St George-Hyslop, P.H., Tanzi, R.E., Polinsky, R.J., Haines, J.L., *et al.* (1987), 'The genetic defect causing familial Alzheimer's disease maps on chromosome 21', *Science, 235:* 885-890.

Stokes, G. & Holden, U. (1993), 'Dementia: Causes & Clinical Syndromes' in Stokes, G. & Goudie, F. (eds), *Working with Dementia*, Bicester: Winslow Press.

Sylvester, P.E. (1984), 'Ageing in the Mentally Retarded' in Dobbing, J., Clarke, A.D.B., Corbett, J.A., Hogg, J. & Robinson, R.O. (eds), *Scientific Studies in Mental Retardation*, London: Royal Society of Medicine/Macmillan, 259-277.

Tangye, S.R. (1979), 'The EEG and incidence of epilepsy in Down's syndrome', *Journal of Mental Deficiency Research, 23:* 17-24.

Thompson, S.B.N. (1993a), 'Down's syndrome and Alzheimer's disease', Therapy Weekly, 19; 21: 8.

Thompson, S.B.N. (1993b), *'Eating Disorders: A Guide for Health Professionals'*, London: Chapman & Hall.

Thompson, S.B.N., North, N. & Pentland, B. (1992), 'Clinical management of a man with complex partial seizures and a severe head injury', *Brain Injury, 7;* 3: 257-262.

Tomlinson, B.E., Blessed, G. & Roth, M. (1970), 'Observations on the brains of demented old people', *Journal of the Neurological Sciences, 11:* 205-242.

Veall, R.M. (1974), 'The prevalence of epilepsy among mongols related to age', *Journal of Mental Deficiency Research, 19:* 99-106.

Verhaart, W.J.C. & Jelgersma, H.C. (1952), 'Early senile dementia in mongolian idiocy. Description of a case', *Folia Psychiatrica Neerlandica, 55:* 453-459.

Warren, A.C., Holroyd, S. & Folstein, M.F. (1989), 'Major depression in Down's syndrome', *British Journal of Psychiatry, 155;* 202-205.

Wechsler, D. (1981), *'Manual for the Wechsler Adult Intelligence Scale – Revised'*, New York: The Psychological Corporation.

Whalley, L.J., Carothers, A.D., Collyer, S., De May, R., *et al.* (1982), 'A study of familial factors in Alzheimer's disease', *British Journal of Psychiatry, 140:* 49-56.

Wisniewski, K.E., French, J.H., Rosen, L.J., Koziowski, P.B., *et al.* (1983), 'Basal ganglia calcification in Down's syndrome - another manifestation of premature ageing', *Annals of the New York Academy of Sciences, 396:* 179-189.

Young, E.C. & Kramer, B.M. (1991), 'Characteristics of age-related language decline in adults with Down's syndrome', *Mental Retardation, 29;* 2: 75-79.

Chapter 4

RATE OF DECLINE IN SOCIAL AND COGNITIVE ABILITIES IN DEMENTING INDIVIDUALS WITH DOWN'S SYNDROME AND OTHER LEARNING DISABILITIES

S.B.N. Thompson

Clinical Gerontologist (2003), 26; 3/4: 145-54.

ABSTRACT

Sixteen clients with learning disabilities, 8 Down's syndrome (DS), 8 non-Down's syndrome (NDS), diagnosed with dementia, were assessed using specially selected neuropsychological assessment tools at two time points separated by six months.

1. DS individuals showed decline in cognitive abilities ($p < .005$; 1-tailed) (hypothesis 1);
2. NDS showed decline in cognitive abilities ($p < .01$; 1-tailed; $p < .005$; 1-tailed) (hypothesis 2);
3. DS individuals showed decline in social abilities ($p < .005$; 1-tailed) (hypothesis 3), but the NDS did not (hypothesis 4);
4. No significant differences were found in the ability score changes of the two groups (hypotheses 5 & 6).

Findings (1-3) support previous studies with participants over 12 months. Hence, the rate of change in these abilities is faster than previously suspected.

INTRODUCTION

In the past, there have been numerous definitions of dementia dependent upon the viewpoint of the person proposing the definition. However, the definition of dementia currently and generally accepted by clinicians is that outlined in DSM-IV (APA, 1994). The salient points of this

definition are that both memory and other cognitive functions should be impaired.

Several studies in the last decade have sought to measure the symptoms that are characteristic of dementia (eg Nagy, *et al.*, 1998; Schmand, *et al.*, 1998). The problems of a differential diagnosis, particularly distinguishing between depression and dementia, are well documented (Thompson, 1997). However, diagnosing dementia in people with learning disabilities can be a particularly difficult process (Deb, 1997; Deb & Braganza, 1999), mainly because of their poor intellectual development and the fact that many assessment tools have floor and ceiling effects for this population (Thompson, 1999a).

Several researchers have attempted to identify dementia in people with learning disabilities using rating scales (Evenhuis, 1996) and direct assessment tools (Hon, *et al.*, 1999; Burt & Aylward, 2000; Thompson, 1999b; 2000). Despite these attempts, still little is known of the differences between dementing individuals with Down's syndrome (DS) and those with other types of learning disabilities. In particular, there is a lack of detailed knowledge of the social and cognitive changes in these populations over time. This study aims to corroborate previous findings

drawn from a further population of DS and other learning disabilities over a shorter time span.

In 1999b, Thompson reported score changes over twelve months, reflecting the social abilities of 20 DS participants which were significantly greater ($p < .002$) than those of the 21 non-Down's syndrome (NDS) learning disability sample group. Findings were explained in terms of poor language abilities in the DS people generally, and the link between declining social abilities and dementia. Participants were assessed using the Dementia Questionnaire for Mentally Retarded Persons (DMR) (Evenhuis, 1992a,b; 1996) to measure cognitive and social abilities, and the Middlesex Elderly Assessment of Mental State (MEAMS) (Golding, 1989) to measure cognitive functioning. Other tools measured intelligence quotient, daily living care skills and anxiety and depression.

Thompson (2000), using the same assessment tools, found a weak linear association ($p < .004$; $r = .625$; 2-tailed) between cognitive performance and social abilities in the DS sample. Scores on a modified version of the Hospital Anxiety and Depression Scale (Zigmond & Snaith, 1983) were found to be in the normal or borderline range. Hence, it was unlikely that either levels of anxiety or depression had affected

participants' performances significantly. Thompson (2000) discussed these findings in relation to Baddeley's working memory model and the Central Executive System theory (Baddeley, *et al.*, 1991; Baddeley, 1992).

These various studies show changes in scores in social and cognitive abilities over a twelve-month period. Thompson (1994) investigated these changes over six and twelve months, finding similar results. However, the sample size was limited to 8 DS, and no NDS participants. This study further examines these changes using some of the same assessment tools but over 6 months and with a larger sample drawn from a DS and a NDS population.

METHOD

Rationale and Hypotheses

The link between people who have the neuropathology of DS and their development of clinical signs associated with Alzheimer's-type dementia has been well documented (Oliver, 1999; Oliver & Holland, 1986;

Thompson, 1997; 2000). Less is known about the effects of dementia on non-DS persons with learning disabilities and whether there is any difference when compared with DS individuals. However, there is growing evidence to suggest that cognitive and social abilities decline with the onset and progress of dementia and that changes in these abilities are different between DS and NDS groups (Thompson, 1999; 2000).

Hypothesis 1: Do cognitive abilities decline over time in DS?

Hypothesis 2: Do cognitive abilities decline over time in NDS?

Hypothesis 3: Do social abilities decline over time in DS?

Hypothesis 4: Do social abilities decline over time in NDS?

Hypothesis 5: Do people with DS show a greater decline in cognitive abilities with age, compared with NDS?

Hypothesis 6: Do people with DS show a greater decline in social abilities with age, compared with NDS?

Materials

Wechsler Adult Intelligence Scale-Revised (WAIS-R) (Wechsler, 1981); Middlesex Elderly Assessment of Mental State (MEAMS); Dementia Questionnaire for Mentally Retarded Persons (DMR); Hospital Anxiety and Depression Scale (HADS).

Sample

Clients were randomly selected from the consultant psychiatrist's list of people from Southern regions of the United Kingdom. All clients agreed to participate in the study and were diagnosed by the consultant psychiatrist as having either Down's syndrome (and allocated to Group 1) or another learning disability (Group 2), ie prenatal abnormality/illness such as rubella or meningitis; perinatal trauma such as anoxia or cerebral palsy; or metabolic such as phenylketonuria. None of the clients had a genetically acquired learning disability.

(i) 'Down's Syndrome' (DS) - Group 1

Three male and 5 female clients with an average age of 49.38 years (sd = 8.07) and range of 35-56 years. Clients were found to have an average full-scale IQ on the WAIS-R of 53.63 (sd = 4.84) and range of 48-63 IQ points.

(ii) 'Non-Down's Syndrome' (NDS) - Group 2

Four male and 4 female clients with an average age of 55.13 (sd = 6.45) years and range of 45-66 years. Clients were found to have an average full-scale IQ on the WAIS-R of 60.00 (sd = 5.53) and range of 51-68 IQ points.

Controls

Each client was his/her own control. Individual's scores obtained at the first assessment were compared with their own scores at the second assessment and then compared between the two sample groups.

Procedure

Permission and approval for this study was granted by the local ethics committees. Consent was obtained from those people in charge of the care establishments approached as well as from individual clients. The following summarises information obtained from each assessment:

First Assessment

1. DMR - sum of cognitive scores and sum of social scores;
2. HADS - total score for anxiety and total score for depression;
3. WAIS-R - verbal IQ, performance IQ and full-scale IQ;
4. MEAMS - total score and number of subtests passed;
5. Biographical/medical details.

Second Assessment (at 6 months)

1. DMR - sum of cognitive scores and sum of social scores;
2. HADS - total score for anxiety and total score for depression;

3. MEAMS (version B) - total score and number of subtests passed.

The WAIS-R was not repeated at assessment 2 in order to avoid the possibility of practice effects.

RESULTS

Test scores obtained for each group of clients were compared. No statistically significant differences were found between the two groups for the WAIS-R Full-scale IQ thus confirming the similarity of ability profiles of the two groups. However, there was a significant difference between the ages (T=12; U=13; $p < .025$; 1-tailed), indicating that the NDS group were significantly older than the DS group. This may reflect the population from which the sample was drawn; subjective analysis revealed there was a tendency for younger people with DS and dementia to be admitted to local care establishments and this was also found to be the case in referrals to clinical psychology departments in the region.

Test scores obtained at assessments 1 and 2 were compared firstly, within groups. For the DS clients (Table 1), DMR Sum of Social Scores and the MEAMS Number of Subtests passed were both found to be significant at the same level (T=0; W=0; $p < .005$; 1-tailed), indicating that there was a decline in social ability scores and also in cognitive functioning subtests passed over the 6-month time interval. Neither anxiety nor depression scores of the HADS differed significantly between assessments 1 and 2.

For the non-DS clients (Table 2), the MEAMS Number of Subtests passed was found to be significant (T=0; W=0; $p < .005$; 1-tailed) and also the MEAMS Total scores (T=1; W=2; $p < .01$; 1-tailed), indicating a decline in cognitive functioning subtests passed over the 6-month time interval as well as the overall scores for cognitive functioning. Neither anxiety nor depression scores of the HADS were significant between assessments.

Comparing measures between the two sample groups (Table 3), no significant evidence was found for any of the measures.

TABLE 1: Group 1: Comparison of test measures between assessments 1 and 2

Variable	Mean	SD	T	W	p
DMR (SCS)	4.13	3.31	9.5	6	ns
DMR (SOS)	6.50	7.69	0.0	0	<.005 (1-tailed)
MEAMS Total	4.00	3.46	13.5	6	ns
MEAMS Sub.	0.63	1.41	0.0	0	<.005 (1-tailed)
HADS Anxiety	1.88	1.64	8.0	6	ns
HADS Depression	0.75	1.16	15.0	6	ns

Key: DMR = Dementia Questionnaire for Mentally Retarded Persons
SCS (SOS) = Sum of cognitive (social) scores of DMR
MEAMS Sub. = Middlesex Elderly Assessment of Mental State number of subtests passed
HADS = Hospital Anxiety and Depression Scale
SD = Standard Deviation
T = Smaller sum of ranks
W = Wilcoxon Signed-Ranks Test value
ns = Not significant

TABLE 2: Group 2: Comparison of test measures between assessments 1 and 2

Variable	Mean	SD	T	W	p
DMR (SCS)	1.50	1.85	17	6	ns
DMR (SOS)	2.25		2.12	8	6 (ns)
MEAMS Total	2.00	1.85	1	2	<.01 (1-tailed)
MEAMS Sub.	0.00	0.00	0	0	<.005 (1-tailed)
HADS Anxiety	0.63	0.74	14	6	ns
HADS Depression	0.63	0.52	12	6	ns

Key: DMR = Dementia Questionnaire for Mentally Retarded Persons
SCS (SOS) = Sum of cognitive (social) scores of DMR
MEAMS Sub. = Middlesex Elderly Assessment of Mental State number of subtests passed
HADS = Hospital Anxiety and Depression Scale
SD = Standard Deviation
T = Smaller sum of ranks
W = Wilcoxon Signed-Ranks Test value
ns = Not significant

TABLE 3: Comparison of test measures between Groups 1 and 2 for assessments 1 and 2

Variable	T	U	p
DMR (SCS)	15.5	15	ns
DMR (SOS)	19.0	15	ns
MEAMS Total	21.5	15	ns
MEAMS Sub.	24.0	15	ns
HADS Anxiety	48.0	15	ns
HADS Depression	29.0	15	ns

Key: DMR = Dementia Questionnaire for Mentally Retarded Persons
SCS (SOS) = Sum of cognitive (social) scores of DMR
MEAMS Sub. = Middlesex Elderly Assessment of Mental State number of subtests passed
HADS = Hospital Anxiety and Depression Scale
T = Smaller sum of ranks
U = Mann-Whitney U Test value
ns = Not significant

DISCUSSION

Past studies have found that over a twelve-month period, people with learning disabilities and dementia decline in their cognitive abilities with age (Thompson, 2000). Over six months, using the same assessment tools, this finding was supported using a sample drawn from a different population.

For the DS group, changes in the sum of social scores of the DMR were found between assessment 1 and 2 ($p < .005$; 1-tailed) (hypothesis 3). These scores were significantly lower after the 6-month interval. The number of subtests passed on the MEAMS were significantly less ($p < .005$; 1-tailed) after this interval. (hypothesis 1). For the NDS group, total scores on the MEAMS were significantly lower at assessment 2 ($p < .01$; 1-tailed), and there were less subtests passed on the MEAMS ($p < .005$; 1-tailed). These findings support hypotheses 2.

Score changes in the sum of social scores (DMR) for the DS group were significantly lower ($p < .005$; 1-tailed) at the second assessment. This finding supports hypothesis 3 and further supports work of past studies (Thompson, 1999b) in which participants were measured over a longer, twelve-month interval. However, there was no significant evidence for

such changes in the NDS group; thus hypothesis 4 is refuted. This finding may be due to the short interval between assessments, ie changes were seen over 12 months in past studies.

Comparing findings between the two groups revealed no significant differences; hence, hypotheses 5 and 6 were refuted. Again, this may be due to the shorter interval between assessments.

The results of this study are interesting for a number of reasons. Firstly, they support previous findings of a decline in cognitive abilities regardless of the type of learning disability. Secondly, the time span of this decline may be as short as 6 months. Clearly, this has implications for future care of dementing individuals with learning disabilities, ie there is no luxury of time in treating such patients since deterioration may be relatively fast. Finally, the cohort failed to show a difference between the two groups. This cannot be explained by the significant difference in ages of the two groups because one would expect older dementing individuals to decline more rapidly or at least, show greater changes over time, as evidenced by previous studies (Crayton & Oliver, 1993; Thompson, 1999b; 2000). It may be that the differences found in previous studies over 12 months are simply not large enough to be significant between groups after just 6 months. Evidently, there is a need for further

investigation into the rate of change in cognitive and social abilities in dementing individuals with learning disabilities.

REFERENCES

APA - American Psychiatric Association (1994*), 'Diagnostic and Statistical Manual of Mental Disorders (DSM-IV). 4th ed'*, Washington DC: American Psychiatric Association, 139-143.

Arendt, T. (2001), 'Disturbance of neuronal plasticity is a critical pathologic event in Alzheimer's disease', *International Journal of Developmental Neuroscience, 19:* 231-245.

Baddeley, A.D. (1992), 'Working memory', *Science, 255:* 556-559.

Baddeley, A.D., Bressi, S., Della Sala, S., Logie, R., , *et al.*. (1991), 'The decline of working memory in Alzheimer's disease', *Brain, 114;* 2521-2542.

Bigler, E.D. (1992), 'The neurobiology and neuropsychology of adult learning disorders', *Journal of Learning Disability, 25;* 8: 488-506.

Crayton, L. & Oliver, C. (1993), 'Assessment of Cognitive Functioning in Persons with Down's Syndrome who develop Alzheimer's Disease'

in Berg, J., Karlinsky, H. & Holland, A. (eds), *Down's Syndrome and Alzheimer's Disease*, Oxford University Press.

Deb, S. (1997), 'Structural neuroimaging in learning disability'. *British Journal of Psychiatry, 171:* 417-419.

Deb, S. & Braganza, J. (1999), 'Comparison of rating scales for the diagnosis of dementia in adults with Down's syndrome', *Journal of Intellectual Disability Research, 43 (Part 5):* 400-407.

Evenhuis, H.M. (1992a), *'Provisional Manual of the Dementia Questionnaire for Mentally Retarded Persons (DMR)'*, Zwammerdam: Hooge Burch.

Evenhuis, H.M. (1992b), 'Evaluation of a screening instrument for dementia in ageing mentally retarded persons', *Journal of Intellectual Disability Research, 36:* 337-347.

Evenhuis, H.M. (1996), 'Further evaluation of the Dementia Questionnaire for Persons with Mental Retardation (DMR)', *Journal of Intellectual Disability Research, 40;* 4: 369-373.

Golding, E. (1989), *'The Middlesex Elderly Assessment of Mental State'*, Bury St Edmunds: Thames Valley Test Company.

Nagy, Z., Vatter-Bittner, B., Braak, H., Braak, E., Yilmazer, D.M., Schultz, C. & Hanke, J. (1998), 'Staging of Alzheimer-type

pathology: An interater-infrarater study', *Dementia & Geriatric Cognitive Disorders, 8;* 4: 248-251.

Oliver, C. (1999), 'Perspectives on Assessment' in Janicki, M.P., Dalton, A.J. (eds), *Dementia, Aging, and Intellectual Disabilities: A Handbook*, Brunner/Mazel: Philadelphia, PA, 123-140.

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

Schmand, B., Geerlings, M.I., Jonker, C. & Lindeboom, J. (1998), 'Reading ability as an estimator of premorboid intelligence: Does it remain stable in emergent dementia?' *Journal of Clinical & Experimental Neuropsychology, 20;* 1: 42-51.

Thompson, S.B.N. (1997), '*Dementia: A Guide for Health Care Professionals'*. Aldershot: Arena.

Thompson, S.B.N. (1999a), 'Assessing dementia in people with learning disabilities for cognitive rehabilitation', *Journal of Cognitive Rehabilitation, 17;* 3: 14-20.

Thompson, S.B.N. (1999b), 'Examining dementia in Down's syndrome (DS): Decline in social abilities in DS compared with other learning disabilities', *Clinical Gerontologist, 20;* 3: 23-44.

Thompson, S.B.N. (1994), 'A neuropsychological test battery for

identifying dementia in people with learning disabilities', *British Journal of Developmental Disabilities, XL;* 2 (79):135-142.

Thompson, S.B.N. (2000), 'The Central Executive System in people with Down's syndrome and dementia', *Clinical Gerontologist, 21;* 3: 3-32.

Wechsler, D. (1981), *'Manual for the Wechsler Adult Intelligence Scale – Revised'*, New York: The Psychological Corporation.

Zigmond, A.S. & Snaith, R.P. (1983), 'The Hospital Anxiety and Depression Scale', *Acta Psychiatrica Scandinavica, 67:* 361-370.

Chapter 5

THE CENTRAL EXECUTIVE SYSTEM IN PEOPLE WITH DOWN'S SYNDROME AND DEMENTIA

S.B.N. Thompson

Clinical Gerontologist (2000), 21; 3: 3-32.

ABSTRACT

Forty-one clients with learning disabilities (20 Down's syndrome, 21 non-Down's syndrome) were assessed using specially selected neuropsychological assessment tools at two time points separated by twelve months. Evidence for hypothesis 1 suggested that people with Down's syndrome show a greater decline in cognitive abilities with age, compared with other groups of people with learning disabilities. A weak linear association ($p < .004$; $r = .625$; 2-tailed) between cognitive

performance and social abilities was also found for the Down's syndrome clients, supporting hypothesis 2. Findings were explained in terms of the link between declining cognitive abilities and dementia and the documented evidence of language deficits in Down's syndrome.

INTRODUCTION

Normal ageing brings with it certain changes to the higher mental or 'cognitive' functions (Allen, et al., 1997). Memory can also be affected (Craik, 1994), sometimes because the individual has failed to receive information correctly, or sometimes because it can no longer be effectively encoded or stored (Nyberg, et al., 1996). The effect of ageing on memory is very often one of the first of the cognitive functions to be noticed by others and can cause considerable distress to the individual and to relatives, close friends and carers. Deterioration in memory functioning is characteristic of dementia (Mitrushina, *et al.*, 1995) but it can also indicate other dysfunctions which should always be considered in any assessment.

Structural changes to the brain give rise to cognitive changes (Daigneault & Braun, 1993) that may be seen by others observing the individual. Anterograde memory functioning (often used to describe the every day memory functioning following an insult to the brain) has been attributed to the mammillary bodies deep within the brain structure. Following head injury, stroke or dementia, these tiny structures may become damaged (Kapur, et al., 1996) leading to loss of memory function. In normal ageing, the brain undergoes several structural changes including decreasing in size, flattening of the surface, and increasing amounts of intracranial space (Jernigan, et al., 1980). Other microscopic and biochemical changes occur as well as changes to the electrical activity (electrophysiological changes) within the brain (Zatz, et al., 1982). Verbal skills, particularly the well-learned skills of reading, writing, vocabulary, and word usage, tend to be maintained (Botwinick, 1977); and the general intellectual status of healthy older people, as measured by neuropsychological tests, tends to remain within normal limits through the eighth decade (Benton, et al., 1981).

Arithmetic ability is also generally stable among older people (Kramer & Jarvik, 1979). Arithmetic and memory tests that show decreased performance in older people, for example, Digits Backward of the

Wechsler Adult Intelligence Scale-Revised (Wechsler, 1981), tend to reflect impaired concentration and mental tracking rather than decreased cognitive functioning (Parkin, *et al.*, 1995). However, a normal tendency for digit and letter span to be a little longer in the auditory than visual modality appears to increase with age (Kramer & Jarvik, 1979). Contrary to conventional belief, normal ageing processes do not affect the immediate memory span in older people (Nyberg, *et al.*, 1996).

Normal intellectual decline associated with old age shows up most strikingly in four areas of intellectual activity; these can be summarized as follows:

1. The primary, or working, memory capacity of intact older people differs little from that of younger adults (Erickson, 1978) except when the amount of material to be remembered exceeds the normal primary storage capacity of six or seven items, as in tests of supraspan (Craik, 1977). Older people use less effective learning procedures-less elaborative encoding (Fisk, *et al.*, 1997) and tend to show a greater differential between recall and recognition of learned material, particularly when the recognition tasks are easy (Botwinick & Storandt, 1974). Contrary to studies

that indicate a progressive loss in recall of public events (Squire, 1974), Botwinick and Storandt (1980) reported that memory for remote events does not appear to change with the passage of time.

2. Diminished ability for abstract and complex conceptualization typifies the intellectual functioning of older people (Botwinick, 1977). The more meaningful and concrete the presentation of reasoning problem, the greater is the likelihood that people will succeed at it (Botwinick, 1978).

3. Mental inflexibility, appearing as difficulty in adapting to new situations, solving novel problems, or changing mental set, characterizes intellectual performance failures of older age (Schaie, 1958). Some authors have suggested that apparent intellectual slowness in solving problems may be due to serial versus parallel processing. Evidence for slower serial processing was found in tests of older people as compared with younger participants (Ellis, *et al.*, 1996).

4. General behavioural slowing (Swearer & Kane, 1996) is a predominant characteristic of ageing that affects perceptual (Earles & Salthouse, 1995), cognitive (Thomas, *et al.*, 1977), and memory functions (Nyberg et al., 1996) as well as psychomotor activity (Benton, 1977; Welford, 1977). Accurate evaluation of an older person's poor performance on any timed test must depend on careful observation and analysis of the effect of time limits on the scores, for the score alone will tell little about the effects of slowing per se (Lezak, 1995).

There are of course several other conditions that might be confused with a diagnosis of dementia in older people. Some of these include paraphrenia, often defined as 'schizophrenia of late life', alcohol-related problems (such as Korsakoff's psychosis), and Parkinson's disease, the most common of which occurs in one or two people in every thousand (Figure 1) (Goudie, 1993).

FIGURE 1: Similarities and differences between dementia and other physical and psychological problems (adapted from Goudie, 1993)

Problem	Similarities to dementia	Differences from dementia
Acute confusional state	Disorientation, poor concentration, self-neglect	Occurs rapidly, worse at night; disappears after underlying causes treated; clouding of consciousness
Depression	Poor concentration; slowness; non-responsiveness	Answers which but 'do not know' is frequent response
Anxiety	Inability to carry out day-to-day tasks because of agitation, catastrophic reaction - total failure to cope	No confabulation; insight into impaired functioning; when stressors minim-ized, ability is as normal
Paraphrenia	Misinterpretation of actions and state-ments, self-neglect	Components of behaviour unimpaired, no missing out of steps in a task even if reasoning seems bizarre; hallucinations

Normal Memory

Memory failure is a common and significant problem in dementia, hence it is important to first assess the extent to which it is a problem and for whom the problem is an obstacle.

A distinction is commonly drawn between a short-term memory system of limited capacity, a few items at most, and a storage of seconds, and a long-term system of perhaps limitless capacity and indefinite storage time (Atkinson & Shiffrin, 1968). Short-term memory, now elaborated into the concept of working memory (Baddeley, *et al.*, 1995), is the system which enables a new telephone number to be remembered while dialling it-so long as there is no distraction. Long-term memory allows one to remember a familiar telephone number from day to day and year to year (Collerton, 1993). This terminology differs from commonplace use where the short-term memory is taken to be the memory for the preceding hours, days or months, and long-term memory for many years in the past.

It is widely accepted that different types of knowledge appear to be stored differently, so that, for example, knowing what a person ate for lunch (episodic memory) would be stored differently from knowing the

word 'lunch' means a mid-day meal (semantic memory). Cohen and Squire (1981) have subsumed these terms under 'declarative memory' and reserve a further definition, termed 'procedural memory' for skills and routines including some types of sensory memory (eg knowing how to ride a motorbike is a procedural memory, knowing how the engine works is declarative). These functional definitions of memory have practical applications for therapists and are also more simplistic than earlier definitions.

DEMENTIA AND THE CENTRAL EXECUTIVE SYSTEM

The definition of dementia generally accepted by clinical psychologists and psychiatrists is that outlined in DSM-IV (APA, 1994). In summary, it states that for a diagnosis of dementia, there should be demonstrable evidence of impairment in short-term and long-term memory. Impairment in short-term memory (ie inability to learn new information) may be indicated by an inability to remember three objects after five minutes. Long-term memory impairment (ie inability to remember information that was known in the past) may be indicated by an inability

to remember past personal information (eg what happened yesterday; birthplace; occupation) or facts of common knowledge (eg past Prime Ministers; well known dates).

Dementia is commonly misunderstood to be a disease when in fact it may be the result of a number of factors, and in some instances it may be reversible. Stokes and Holden (1993) have described 'primary dementia' as an extensive, organic impairment of intellect memory and personality. It occurs in the absence of clouding of unconsciousness (ie without drowsiness) which is acquired, irreversible and progressive.

The single most common form of dementia is Alzheimer's disease which accounts for more than 50 per cent of cases of dementia in those over the age of 65 (Katzman, 1976). Typically, the onset is from 40 years of age onwards with insidious degeneration until death at about two to five years following onset (Jorm, 1990).

Each person with Alzheimer's disease will vary slightly in presentation according to personality. Emotional, behavioural and cognitive changes will also vary, but generally accepted by clinicians and researchers is a stage model which describes broad characteristics (Reisberg, 1983). In the first phase, the 'forgetfulness phase', there is usually difficulty in recalling recent events, and a tendency to forget where objects have been

placed. Names of people and places, previously familiar, may be poorly recalled and a general disorientation persists and poor short-term memory. Abstract thinking, inability to concentrate on tasks and a marked lack of curiosity are also typical presentations. There may also be emotional changes such as anxiety and irritability and the 'new' or unexpected will be feared or disliked. Denial is also sometimes seen in presentation of people with Alzheimer's disease.

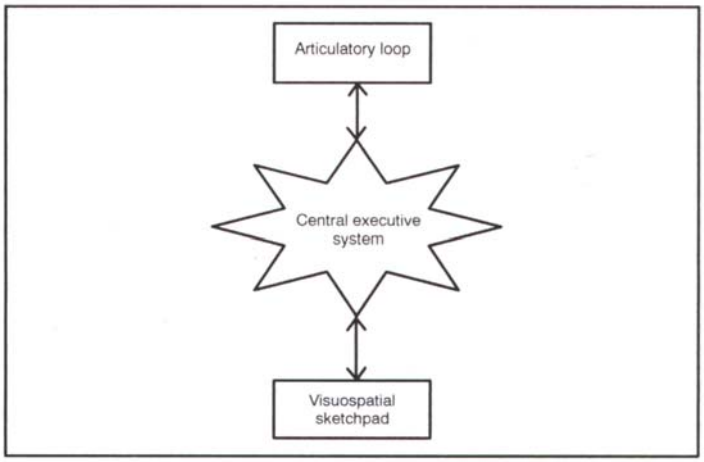
The second recognised phase is known as the 'confusional phase'. Increasingly poor attention span and a decline in generalised intellectual performance is seen with a deteriorating memory. Disorientation in place, word-finding difficulty and other changes to speech may be seen. Complex tasks are performed with difficulty, sometimes in a clumsy or inaccurate manner and often the skills the person learned last will be lost first. Hence the skills necessary for social independence and vocational skills are usually the first skills to be reduced or lost completely. Together with failing memory comes the concealment of these deficits by rationalising or confabulating events (ie providing an imaginary account of events or actions). Lack of interest in news and surroundings follows relatively quickly and can be extremely distressing to family and friends.

The third phase, the 'dementia phase', is characterized by a lack of purpose in the person's behaviour which appears disjointed and sometimes bizarre. Remaining intellectual and self-care abilities require constant supervision as people in this phase undergo further deterioration in memory capacity, calculating ability (dyscalculia) and aspects of language are severely affected and eventually lost. Constant assistance is required for self-care skills such as grooming, dressing, toileting and for feeding. A progressive physical wasting can also be seen which will mean help with walking. Sometimes one or two years of life will follow in an almost vegetative state until death.

In normal functioning, several types of information may be processed at once, possibly from a variety of sources. An example of this might be holding a conversation or driving a car. These types of activities break down relatively rapidly in patients with dementia, even at the early stages. Alberoni et al. (1992) reported that Alzheimer-type patients are particularly handicapped in keeping track of conversations involving several people. They also often have problems in remembering who said 'what' and 'when'.

This type of mental activity is successfully characterised by the Working Memory Model (Fig 2) developed by Baddeley and colleagues

FIGURE 2: Adapted from Baddeley's (1992) Working Memory Model



Source: Morris, R.G. (1994). 'Recent developments in the neurospsychology of dementia. *International Review of Psychiatry, 6:* 85-107.

(Baddeley, 1986; 1992; Baddeley & Hitch, 1974). The model proposes a Central Executive System (CES) which functions to co-ordinate and schedule mental operations including the processing and immediate storage of information. This incorporates the notion of a 'scratch pad' system which holds and manipulates information simultaneously. The CES has limited resources, hence the decline in performance associated with trying to combine two attentionally demanding mental operations. A cluster of peripheral systems support the functioning of the CES. This includes a specialised Articulatory Loop System (ALS), responsible for recycling verbally encodable information, such as when a person tries to keep in mind a telephone number for a short period or understand a grammatically complicated sentence.

In the visuospatial domain, there is the visuospatial scratchpad (VSSP) which maintains visuospatial imagery. These components of the model are interacting continuously with the CES devoting varying degrees of processing resources to each component depending on the task. This is not the only model explaining sequencing and executive phenomena (Shallice, 1988; Furster, 1993), but it has provided a useful framework for studying cognitive impairment in dementia which has in

part been characterised as a dysexecutive syndrome (Morris, 1986; Baddeley, *et al.*, 1986; Becker, 1988).

The peripheral systems of the Working Memory Model (Baddeley, 1992) are coordinated by the CES. The most extensively studied is the ALS which recycles verbal material in a relatively automatic fashion. For example, when remembering a string of digits, as in the digit span test of the Wechsler Adult Intelligence Scale Revised (Wechsler, 1981), the material is cycled continuously through the ALS to keep it in memory. A series of studies indicate that this system is unimpaired in Alzheimer's disease (Morris, 1984; 1987a,b; 1992). The evidence comes from experiments where patients are required to recall short lists of digits or words but the functioning of the ALS is suppressed by requiring them to mouth an irrelevant word (such as the word 'the'), effectively blocking articulatory rehearsal. 'Articulatory suppression' of this sort was found to have quantitatively the same effect on digits or words recall in Alzheimer's disease patients as in normal subjects. Thus, the equivalent loss of function without articulatory rehearsal shows the potency of the system to recycle verbal material is undiminished in Alzheimer's disease (Morris, 1984; 1987a,b; 1992).

The visual counterpart of the ALS is the visuospatial sketchpad which functions to retain visuospatial memory in immediate memory (Baddeley, 1992). For example, if a person's attention is diverted momentarily from a visual scene, they will still maintain the memory of that scene for a short period. This ability helps the person maintain a sense of visual continuity when, for example, moving around the room. Although this ability has not been investigated extensively, there is evidence for significant impairment in visuospatial memory in Alzheimer's disease (Morris, 1994a). Firstly, patients with Alzheimer's disease have difficulty in a visual analogue of verbal span – the patient observes the experimenter tapping out a sequence on an array of nine blocks (or printed coloured squares as in the Wechsler Memory Scale Revised-Wechsler, 1988) in front of them and then has to immediately repeat the sequence of taps from memory. This block span performance has been found to be impaired consistently (Spinnler, *et al.*, 1988). Secondly, there is a measure known as the 'delayed matching to sample task' where an item is shown to the patient followed by a short delay followed by the same item with another or several others. The patient is required to pick out the item seen before. By varying the delay it is possible to see how fast memory for the item decays. Sahakian, *et al.* (1988) used a version in

which a pattern was shown on a computer screen and the patient had to identify the pattern from four others. Early Alzheimer's disease patients were found to show more rapid forgetting than normal older controls. A later study by Money, *et al.* (1992) found a similar impairment using filled circles of different sizes, but Alzheimer's disease patients were impaired when there was no delay between presentation and choice and showed the same rate of forgetting as the controls. Both studies, therefore, showed a substantial impairment in the performance of Alzheimer's disease patients.

Baddeley et al. (1991) favours a localisationalist view; impairment in Alzheimer-type patients may be explained in terms of dysexecutive syndrome and frontal lobe dysfunction. This links the notion of the CES to the Norman and Shallice (1986) model of attentional control which assumes that most ongoing actions are controlled by establishing routines. The routines or 'schemas' are mutually inhibitory and can be triggered by environmental events. Where they are insufficient to generate appropriate activity, a higher-level system, the Supervisory Attentional System (SAS) comes into play which is involved in coping with novel circumstances or problem-solving activity (Morris, 1994b).

Shallice (1988) relates an impairment in the SAS to the difficulties patients with frontal lobe damage have in problem-solving. The conceptual link between the CES and the SAS has been introduced by Baddeley et al. (1991), who suggests that an SAS impairment also results in the reduced capacity to direct and control attentional resources. Thus an impairment in the SAS may be closely analogous to the dysexecutive syndrome seen in Alzheimer's disease. Indeed, Alzheimer's disease patients do have damage to their frontal lobes (Morris, 1994b).

LEARNING DISABILITY

'Learning disability' is a very broad term and has been used to describe people with an intelligence quotient (IQ) below 70. Wechsler (1981) has classified the abilities of groups of people according to IQ. People with learning disabilities commonly may have a range of difficulties which might include approaches to problem-solving, coordination difficulties, problems with speech or comprehension, cognitive delay, or slowness or inability to perform daily routines, such as hygiene or feeding (Thompson, 1993). The range or number of difficulties an individual may

have can be very large or equally, very small. Increasingly, therefore, it has been useful to state a person's abilities rather than emphasising their negative disabilities. With the promotion of community living, definitions of learning disability have come to include the extent of a person's ability to live alone or his or her 'independence'.

Down's Syndrome

Down's Syndrome (DS) is one of the most frequently observed forms of learning disability (Moody & Moody, 1992). A characteristic of DS is the presence of an extra gene on chromosome ('trisomy') 21 (Shermann, *et al.*, 1991). Often the person has developmental delays (Maclean, *et al.*, 1991), sometimes a slightly larger head circumference (Palmer, *et al.*, 1992), and language difficulties, especially with ageing (Young & Kramer, 1991). DS has been the focus of much research and controversy (Barr, 1990); because of increased longevity (Eyman, *et al.*, 1991), individuals with DS are living long enough to be at risk for a host of age-related diseases (Young & Kramer, 1991), for example, pre-senile dementia (Wisniewski, *et al.*, 1983).

Jervis (1948) and Verhaart and Jelgersma (1952) described clinical deterioration associated with Alzheimer-like changes at post mortem in a number of people with DS; subsequent research focused on establishing similarities between the neurofibrillary tangles and granulovascular degeneration characteristic of Alzheimer's disease. By the 1960s the link between the two disorders was clearly established; and it was agreed that all people with DS over the age of 35 have the neuropathological features of Alzheimer's disease (Heston, 1977).

However, there is still debate over whether or not people with DS also present with typical clinical features of Alzheimer's disease (Roper & Williams, 1980), since many such individuals maintain good physical and mental health into the fourth and fifth decades of life (St. Clair & Blackwood, 1985). Interest in the relationship between Alzheimer's and DS has been further stimulated by discoveries localising two distinct Alzheimer's disease markers to chromosome 21. First, the gene encoding the precursor protein which, in some processed form, gives rise to the amyloid deposits in Alzheimer's disease, is found on chromosome 21 (Kang, *et al.*, 1987). Second, a marker has been identified on chromosome 21 which is linked to within 15 centrimorgans (a distance spanning 300-600 genes or 15 million base pairs) of a site associated with

an autosomal dominant form of Alzheimer's disease (St. George-Hyslop, *et al.*, 1987). Arguments in favour of some kind of connection have been advanced for some time (Prosser, 1989), but evidence from some studies also puts doubt on such a definitive connection; for example, it is suggested that the amyloidogenic gene on chromosome 21 is not identical to the Alzheimer's gene despite the widespread presence of amyloid material in the senile plaques and neurofibrillary tangles (Curan & Wattis, 1989). This might imply the picture reflected in DS may be of a different brain process to that evidenced in sufferers of Alzheimer's disease.

The major difficulty in applying dementia diagnostic criteria is the underlying learning disability. As Levinson and colleagues (1955) have commented, there is great variation in the development theories learning disabilities. These hold great restraints on the application and interpretation of conventional test procedures (Atkinson, 1991): failure or poor performance might indicate dementia or an underlying. Clearly, these restrictions would not be necessary if the 'intelligence' of this client population was evenly distributed throughout age groups.

Although a high proportion of individuals with DS develop the neuropathological changes of Alzheimer's disease, only a proportion develop the definite signs of deterioration and have the clinical features

characteristic of the later stages of Alzheimer's disease. It is also often difficult to discriminate between pre- or perinatal brain damage (eg meningitis; anoxia) in association with normal ageing (Gath, 1986) and those considered to be the result of a dementing process (Haxby, 1989). The situation is more complicated in people with a learning disability when there can be other confounding variable such as the long-term effects of institutional living, communication and comprehension difficulties, and the lack of a premorbid intelligence quotient since intellectual deficits may have originated from birth.

The paradox between unequivocal neuropathological findings and limited clinical evidence of dementia, particularly in DS, has been partly resolved by the use of specific neuropsychological assessments to detect age-relted deficits (Miniszek, 1983) but conclusive evidence to distinguish clinical features of dementia from normal ageing in people with DS is still not available.

METHOD

Rationale and Hypotheses

A link has been known for some time between people who have the neuropathology of DS and their subsequent development of clinical signs, associated with Alzheimer's-type dementia. Less is known about the effects of dementia on non-DS persons with learning disabilities and whether there is any difference when compared with DS individuals. However, there is substantial evidence to show that cognitive abilities may decline with the onset and progress of dementia.

Hypothesis 1: Do people with DS show a greater decline in cognitive abilities with age compared with other groups of people with learning disabilities?

Hypothesis 2: Do cognitive abilities decline over time together with social abilities?

Materials

Wechsler Adult Intelligence Scale-Revised (WAIS-R) (Wechsler, 1981); Raven Coloured Progressive Matrices (RCPM) (Raven, *et al.*, 1995); Middlesex Elderly Assessment of Mental State (MEAMS) (Golding, 1989); Hampshire Social Services Assessment (HSSA) (HSS, 1989). (To make the HSSA more user-friendly and less confusing to use, zero scoring items which were indicated by an asterisk in the original version, were changed to a zero and normal typeface headings were changed to bold typeface).

Golding and others have discussed the use of the MEAMS dementia screening tool (Golding, 1989; van Belle, *et al.*, 1990). In the MEAMS manual, Golding states that it systematically surveys the major areas of cognitive performance, namely, orientation, name learning, comprehension, remembering pictures, arithmetic, spatial construction, fragmented letter perception, unusual views, usual views, verbal fluency, and motor perseveration. Score 10-12 are ‘normal’ and scores 8 or 9 are ‘borderline range’. Although she states in her three-level criteria, score performance, she does not report test-retest data using the parallel version

of the test. Hence, the actual amount of discrepancy seems to be very dependent on the population studied (Powell, *et al.*, 1993).

Dementia Questionnaire for Mentally Retarded Persons (DMR) (Evenhuis, 1992a); Hospital Anxiety and Depression Scale (HADS) (Zigmond & Snaith, 1983); information sheets for participants; sheets for the collection of clients' biographical details, and consent forms.

Sample

(i) 'Down's Syndrome' (DS)-Group 1

Eleven male and 9 female clients (DS1-DS20) with an average age of 47.25 years (sd = 1.33) and range of 35-63 years (Table 1) were randomly selected from the consultant psychiatrist's list of people from the South of the United Kingdom. All clients agreed to participate in the study. All clients were diagnosed by the consultant psychiatrist as having Down's Syndrome and were found to have an average full-scale IQ on the WAIS-R of 51.90 (sd = 1.21) and range of 47-63 IQ points. Scores of clients' performances on the RCPM placed the majority of clients in the "less than

TABLE 1: Group 1: Summary of HSSA and WAIS-R results

CLIENTS (m=male; (f=female)	HSSA Score	HSSA Band	AGE	WAIS-R (IQ) Verb.	WAIS-R (IQ) Perf.	WAIS-R (IQ) F-Scale
MEAN	37.83	1.65	47.25	54.75	59.00	51.90
SD	4.86	3.03	1.33	1.32	1.12	1.21

Key: Verb. = Verbal IQ
Perf. = Performance IQ
F-Scale = Fullscale IQ
SD = Standard Deviation

the 5th percentile" (Raven, *et al.*, 1995) band, indicating low intellectual ability.

HSSA scores of the majority of clients (13) fell into band 1, indicating "very minimal support needed (i.e, up to 1 hour in 24, often less)" (HSS, 1989; p.7). Five clients fell into band 2, and two clients fell into band 5; the latter indicating "support needed in all major areas (on average over 7/8 hours in 24). Sleeping-in staff or waking staff needed". Mean band: 1.65; sd: 3.03; range: 1-5.

(i) 'Non-Down's Syndrome' (NDS)-Group 2

Ten male and 11 female clients (NDS1-NDS21) with an average age of 56.43 (sd = 1.21) years and range of 45-68 years (Table 2) were randomly selected from the consultant psychiatrist's lists of people from the South of the United Kingdom. All clients were diagnosed by the consultant psychiatrist as having a learning disability other than DS and fell into the following categories: prenatal abnormality/illness (eg rubella; meningitis); perinatal trauma (eg anoxia; cerebral palsy); or metabolic (eg phenylketonuria). None of the clients had a genetically acquired learning disability.

TABLE 2: Group 2: Summary of HSSA and WAIS-R results

CLIENTS (m=male; (f=female)	HSSA Score	HSSA Band	AGE	WAIS-R (IQ) Verb.	WAIS-R (IQ) Perf.	WAIS-R (IQ) F-Scale
MEAN	55.19	2.29	56.43	61.90	63.43	59.19
SD	2.29	1.75	1.21	1.18	1.14	1.17

Key: Verb. = Verbal IQ
Perf. = Performance IQ
F-Scale = Full-scale IQ
SD = Standard Deviation

All agreed to participate in the study. Clients were found to have an average full-scale IQ on the WAIS-R of 59.19 (sd = 1.17) and range of 51-69 IQ points. Scores of clients' performances on the RCPM placed the majority of clients in the "less than the 5th percentile" band, indicating low intellectual ability; five clients were placed in the higher 25th percentile ability band.

HSSA scores ot ten clients fell into band 3, indicating "support for several aspects of care, supervision and independence training, on average 3 or 4 hours in 24" (HSS, 1989; p. 7). Six clients fell into band 1, four clients fell into band 2, and one client fell into band 4; the latter indicating "support for several aspects of care". Mean band: 2.29; sd: 1.75; range: 1-4.

Controls

Each client was his/her own control. Individual's scores obtained at the first assessment were compared with their own scores at each subsequent assessment. In this way, it was possible to identify those clients who showed probable decline in cognitive abilities, as reflected in their change in scores over time.

Procedure

Permission and approval for this study was granted by the local ethics committees. Consent was obtained from those people in charge of the caring establishments approached as well as from individual clients. The following summarises information obtained from each assessment:

First Assessment

1. HSSA-total score and band reflecting the client's dependence;
2. DMR-sum of cognitive scores and sum of social scores;
3. HADS-total score for anxiety and total score for depression;
4. RCPM-total number of correct responses and percentile;
5. WAIS-R-verbal IQ, performance IQ and full-scale IQ;
6. MEAMS-total score and number of subtests passed;
7. Biographical details of each clients tested.

Second Assessment (at 12 months)

1. DMR-sum of cognitive scores and sum of social scores;

2. HADS-total score for anxiety and total score for depression;
3. RCPM-total number of correct responses and percentile;
4. MEAMS (version B)-total score and number of subtests passed.

RESULTS

Scores obtained using the RCPM, reflected performances below the 5th percentile in all but one case for each client group (DS17 & NDS9), which were both at the 10th percentile. Test scores obtained for each group of clients were compared. No statistically significant differences were found between the measures: Age, WAIS-R, Full-scale IQ and HSSA thus confirming the similarity of ability profiles of the two groups.

Test scores obtained at assessments 1 and 2 were compared. For the DS clients (Table 3), all except the MEAMS Total scores were found to be significant. After controlling for the effects of 'anxiety' and 'depression' by partialling out these HADS scores, the following were found to be significant (Table 4): the 'sum of social scores' of the DMR ($p < .001$, HADS Anxiety out; $p < .003$, HADS Depression out). No significant interaction effects were found.

TABLE 3: Group 1: Comparison of test measures between assessments 1 and 2

Variable	Mean	SD	SE	df	F	p
DMR (SOS)	16.70	9.88	2.21	19	132.229	<.001
DMR (SCS)	3.54	1.66	0.37	19	7.535	<.04
MEAMS Tot.	25.40	15.00	3.35	19	1.823	.202 (ns)
MEAMS Sub.	1.61	0.86	0.19	19	23.202	<.001
HADS Anxiety	1.39	0.24	5.42E-02	19	4.901	<.01
HADS Dep.	1.34	0.86	0.19	19	18.127	<.001

TABLE 4: Group 1: Comparison of test measures between assessments 1 and 2 controlling for 'anxiety' and 'depression'

Variable	Anxiety Out			Depression Out		
	df	F	p	df	F	p
DMR (SOS)	14	160.541	<.001	14	84.416	<.003
DMR (SCS)	15	4.668	.190 (ns)	15	10.648	.089 (ns)
MEAMS Tot.	16	1.192	.519 (ns)	16	1.458	.580 (ns)
MEAMS Sub.	2	58.579	<.001	2	33.184	<.001

Key to Tables 3 & 4:

SD = Standard deviation
SE = Standard error
DMR = Dementia Questionnaire for Mentally Retardred Persons
SOS = Sum of social scores of DMR
SCS = Sum of cognitive scores of DMR
MEAMS Tot. (Sub.) = Middlesex Elderly Assessment of Mental State total scores (subtests passed)
HADS Dep. = Hospital Anxiety and Depression Scale depression score
ns = Not significant

For the non-DS clients (Table 5), all except the 'sum of social scores' of the DMR and Anxiety scores were found to be significant. After partialling out the HADS scores (Table 6), the following were found to be significant: the 'sum of cognitive scores' of the DMR ($p < .05$, HADS Anxiety out); and the MEAMS number of sub-tests passed ($p < .001$), HADS Anxiety out; $p < .001$, HADS Depression out). No significant interaction effects were found.

The decline in cognitive scores on the DMR for the DS group was greater than that of the non-Down's syndrome group ($p < .04$; $df = 1$; $F = 7.630$). These findings support hypothesis 1 which suggests people with DS show a greater decline in cognitive abilities with age.

Examining Linear Relationships Within the Data

For the DS clients, an association was found between the 'sum of social "difference" scores' of the DMR with the MEAMS number of subtests passed "difference" scores.

After controlling for HADS Anxiety and HADS Depression scores, this association remained significant ($r = .522$; $p < .04$; 1-tailed) (Table 7). No statistically significant relationship was found with the 'sum of

TABLE 5: Group 2: Comparison of test measures between assessments 1 and 2

Variable	Mean	SD	SE	df	F	p
DMR (SOS)	14.05	9.34	2.04	20	1.255	.396 (ns)
DMR (SCS)	2.46	1.84	0.40	20	3.388	<.02
MEAMS Tot.	26.62	13.37	2.92	20	4.109	<.05
MEAMS Sub.	1.67	0.61	0.13	20	19.693	<.001
HADS Anxiety	1.32	0.38	8.32E-02	20	0.928	.546 (ns)
HADS Dep.	1.17	0.98	0.21	20	2.978	<.05

Key: SD = Standard Deviation
SE = Standard Error
DMR = Dementia Questionnaire for Mentally Retardred Persons
SOS (SCS) = Sum of social (cognitive) scores of DMR
MEAMS Tot. (Sub.) = Middlesex Elderly Assessment of Mental State total scores (no. subtests passed)
HADS Dep. = Hospital Anxiety and Depression Scale depression score
ns = Not significant

TABLE 6: Group 2: Comparison of test measures between assessments 1 and 2 controlling for 'anxiety' and 'depression'

Variable	Anxiety Out			Depression Out		
	df	F	p	df	F	p
DMR (SOS)	13	2.467	.163 (ns)	13	1.106	.493 (ns)
DMR (SCS)	12	6.008	<.05	12	3.203	.082 (ns)
MEAMS Tot.	14	3.080	.143 (ns)	14	2.743	.170 (ns)
MEAMS Sub.	2	14.366	<.001	2	14.081	<.001

Key to Table 6:

DMR = Dementia Questionnaire for Mentally Retarded Persons
SOS = Sum of social scores of DMR
SCS = Sum of cognitive scores of DMR
MEAMS Tot. (Sub.) = Middlesex Elderly Assessment of Mental State total scores (no. subtests passed)
r = Partial correlation coefficient
ns = Not significant
(Note: Parentheses indicate 1-tailed testing; otherwise 2-tailed testing shown.)
($r = .625$; $p < .004$; 1-tailed).

TABLE 7: Partial Correlation Coefficients for Group 1 before and after controlling for 'anxiety' and 'depression'

Variable	Significance Level					
	Before			After		
	df	r	p	df	r	p
DMR (SOS) difference scores with MEAMS Sub. difference scores	18	0.625	<.004 (<.003)	14	0.522	<.04 (<.02)
DMR (SCS) difference scores with MEAMS Sub. difference scores	6	0.580	.132 (ns)	2	0.934	.066 (ns)

Key to Table 7:

DMR = Dementia Questionnaire for Mentally Retarded Persons
SOS = Sum of social scores of DMR
SCS = Sum of cognitive scores of DMR
MEAMS Tot. (Sub.) = Middlesex Elderly Assessment of Mental State total scores (no. subtests passed)
r = Partial correlation coefficient
ns = Not significant
(Note: Parentheses indicate 1-tailed testing; otherwise 2-tailed testing shown.)
(r = .625; p < .004; 1-tailed).

cognitive "difference" scores. For the non-DS clients, these measures were not found to be significantly associated.

These findings suggest that for the DS clients, there is a weak association between the social ability scores of the DMR with the scores of the MEAMS. This supports hypothesis 2 suggesting that both cognitive abilities (as measured by the MEAMS, for example) and social abilities (as measured by the DMR, for example) decline over time.

DISCUSSION

Social Abilities

On comparing data obtained from both client groups, a statistically significant difference was found in the 'sum of social "difference" scores' of the DMR between assessments 1 and 2 ($p < .002$). Although the majority of HADS scores for clients fell within the 'normal' range (i.e., below 8 points), the effect of controlling for these scores was evidenced when they were partialled out in data analyses: $p < .02$ (after HADS Anxiety was partialled out), and $p < .003$ (after HADS Depression was

partialled out). Therefore, the level of statistical significance of this finding was reduced following controlling for the HADS scores.

It it perhaps not surprising to find that anxiety and depression are related to the level of social abilities. This finding tends to support what is already known about the influence of depression and anxiety levels on clients' abilities (Yapa & Roy, 1990; Agbayewa, *et al.*, 1991) and it is suspected that although the HADS scores for the majority of clients in the study were not in the 'borderline' or 'caseness' range, some level of anxiety and/or depression does seem to affect a person's capabilities and performance in the area of social skills.

In addition, the DS clients showed a greater score difference between assessments 1 and 2 in the 'sum of social scores' than the non-DS clients. It is known that people with DS very often have expressive (Young & Kramer, 1991) and receptive (Hartley, 1982) language difficulties. It is proposed that the differences in scores between people with DS and other learning difficulties may be due partly to the known differences in understanding language in the ability for expressive language. Since language is important in the development of social abilities, this impediment may well account for the difference in social abilities seen between the two client groups.

Cognitive Abilities

On comparing scores obtained on the MEAMS number of subtests passed, 4 of the 20 DS clients showed decline, 7 clients slightly improved, and 9 showed no change in their scores. Of the 21 non-DS clients, 9 showed a decline in scores, 1 slightly improved, and 11 showed no change. A significant score change was found ($p < .001$) in the DS clients suggesting that cognitive abilities co-vary over time.

It may well be that decline in social abilities occurs before decline in cognitive abilities in this population, or possibly that the latter is more subtle and difficult to measure in these clients. A decline in social skills, for example, may be more noticeable and easily assessed because of the person's withdrawal from socialising or because of decreased interaction with care staff. Little is known in the learning disability population about the rate of decline or whether decline in cognitive abilities occurs in the same way as it seems to occur in people without learning disabilities who are dementing; although, rate of cognitive decline with age in people who are not dementing, who have DS or "other" learning disabilities seems to be indistinguishable from one another (Das, *et al.*, 1993).

For the DS group, only two clients (DS11 and DS12) scored above 7 (number of subtest passed) at assessment 1. At retest (assessment 2), no clients scored above 7; hence, according to Golding (1989) this would place all these clients in the third (lowest) category where 'more detailed investigation' is recommended. It is curious to note the slight improvement in some scores though all scores were still below the 7-point cut-off stated by Golding (1989) with a mean score for the "differences" between assessments 1 and 2 (for all DS group data) of only 0.30, and a very large variance (standard deviation = 13.33).

For the non-DS group, the mean of scores for the MEAMS number of subtests passed with higher than for the DS group with a mean for these "difference" scores of – 0.57 and a lower variance and less spread than for the DS group (standard deviation = 3.51). Although no statistical difference was found between the two client groups on the MEAMS number of subtests passed, it would seem that of the two client groups, more clients in the non-DS group showed a deterioration in their performance on the MEAMS than did the DS group. It is possible that cognitive abilities were more developed in the non-DS clients and may have been more prone to the debilitating effects of dementia.

Cognitive impairment is salient in dementia in the non-learning disabled population. In particular, executive phenomena have received recent interest which have led to the development of a new assessment measure, the Behavioural Assessment of the Dysexecutive Syndrome (Wilson, *et al.*, 1996). Shallice (1988) and Furster (1993) have also explained the cognitive impairment in dementia in terms of a failure in sequencing and executive phenomena, such as the ability to organise and plan, an Morris (1984; 1986), and others, have described this is in terms of a 'dysexecutive syndrome'. Yet, there is still a lack of substantive evidence of such familiar impairments in people with learning disabilities who have dementia, although it is thought that people with DS tend to decline cognitively (Das, *et al.*, 1993), but the rate of decline seems to vary considerably. Neither has this been particularly well documented, nor have there been well-matched comparisons with non-DS learning disabled people with a diagnosis of dementia.

The cognitive impairment seen in people with dementia may also be considered in terms of Baddeley's (1986) Central Executive System(CES). A breakdown in one of the supporting systems to the CES, the Articulatory Loop System (ALS), responsible for recycling verbally encoded information, may account for the difficulties that learning

disabled people (particularly DS) tend to have with tasks that require intact verbal memory functioning. Language difficulties also impact on tests of cognitive functioning that require verbal responses, for example, the 'verbal' subtests of the WAIS-R and the world fluency subtest of the MEAMS. However, despite some reports of articulatory rehearsal impairments in Alzheimer's disease (eg Hulme, *et al.*, 1993), impressive evidence comes from 'articulatory suppression' tasks (Morris, 1992) which tend to suggest that perhaps the ALS is not implicated in dementia since, particularly Alzheimer-type patients, perform equally well to normal controls. This finding may lend support to the lack of a significant difference in this study between the two client groups in their MEAMS scores.

In the visuospatial domain, the visuospatial sketchpad (Baddeley & Hitch, 1974; Baddeley, 1986), which maintains visuospatial imagery, is thought to be operative in tasks requiring spatial construction (Sahakian, *et al.*, 1988); to some extent, this applies to the visuospatial subtest of the MEAMS. Visuospatial ability is thought to be significantly impaired in people with dementia; however, it has not been investigated extensively (Morris, 1994a).

Examining Linear Associations Between Measures

It does seem to be the case that cognitive abilities (as measured by the DMR 'sum of cognitive "difference" scores') are not associated in a linear manner with scores on the MEAMS. No significant linear relationship was found for date of either client group. However, a significant (though weak) linear relationship was found between the DMR 'sum of social "difference" scores' with the MEAMS number of subtests passed "difference" scores ($r = .522$; $p < .04$; 1-tailed; after controlling for HADS scores) for the DS group. Hence, it is suggested that if social abilities decline in people with learning disabilities (particularly those with DS) then this decline may correlated with their performance on the MEAMS.

Examination of data for each client group individually, revealed that for the DS group, there was a significant difference between assessments 1 and 2 on the DMR 'sum of social scores' ($p < .001$). A significant difference remained following the partialling out of HADS scores. These scores were not significant for the non-DS group. Hence, this is strong evidence for suggesting that social abilities significantly co-vary over

time in people with DS, thus "partially"supporting hypothesis 2, since evidence was only found for the Down's Syndrome group.

Data from the non-DS clients also supported the view that cognitive abilities (as measured by the DMR 'sum of cognitive scores') decline over time ($p < .05$; HADS Anxiety partialled out). Although, when HADS Depression was partialled out, these scores were not significant. It is possible that some of these clients suffered from mild depression (whether or not as a consequence of dementia) and hence their performance on tests of cognitive abilities were affected.

These data tend to partially support hypothesis 2 in that cognitive abilities seem to co-vary over time for both groups of clients.

Problems of a Differential Diagnosis

In terms of the HADS scores between assessments 1 and 2, it was found that both HADS Anxiety and HADS Depression scores co-varied over time, though more significantly in the DS group. This finding can possibly be explained in terms of people's frustrations and low mood which has been evidenced in the non-learning disability population who are dementing (Lamberty & Bieliauskas, 1993). However, it is not known

whether the presence of depression is more salient in people with DS versus other learning disabilities with or without dementia. However, it is important here to address the problem of a differential diagnosis, particularly between dementia and depression. HADS scores were found to be in the borderline range with the majority falling into the 'normal' category. It is recognized that discriminating between dementia and depression in the non-learning disability population is difficult but not impossible, for example, using the Beck Depression Inventory (Beck & Steer, 1987) in conjunction with measures of social and cognitive abilities and the clinical interview.

However, in the learning disability population, it is increasingly difficult when language and comprehension is limited. Despite these difficulties, it is believed that the modified use of the HADS (in an interview situation) and the fact that third parties (namely carers) also contributed to the data pool (by completing the DMR, for example), helped to strengthen the methodology implemented, and hence produce valid results. In addition, biographical data helped to confirm that none of the clients had been observed to be depressed prior to the study nor did any of them have a known history of depression or low mood swings.

Recently, the link between probable Alzheimer's disease and stroke has been reported (Nash, 1997). Studying nuns from a convent in the United States, Snowdon believes that nuns who suffered a stroke also increased their chance of developing Alzheimer's disease. This supports the view that Alzheimer's disease should perhaps be studied together with vascular disease, particularly transient ischaemic attacks, multi-infarcts and cerebrovascular disease.

CONCLUSIONS

A statistically significant difference ($p < .001$) was found in the MEAMS number of subtests passed (i.e., "cognitive abilities") between assessments for the DS and non-DS clients. This decline was greater in the DS clients. This supports hypothesis 1, which proposed that the DS clients showed a greater decline in cognitive abilities with age, compared with other groups of people with learning difficulties.

Several explanations were proposed for the differences found between the two client groups; namely, the well-documented difficulties with language (comprehension and expression) in people with DS may affect

their social abilities. Also, in comparison with Evenhuis' (1992a,b) samples, decline in social abilities has been found to be associated with a dementing process particularly in people with DS.

Evidence was found to support hypothesis 2 which suggested that cognitive and social changes co-vary over time in these two older populations. Social and cognitive abilities tended to co-vary in the non-DS clients whilst cognitive abilities tended to co-vary in the non-DS clients. These findings were discussed in terms of the 'dysexecutive syndrome' (Shallice, 1988; Furtster, 1993; Wilson, *et al.*, 1996) which suggests that cognitive impairment in dementia occurs because of a failure in sequencing and executive phenomena, such as planning and organising. Impairments possibly due to changes in visuospatial ability were also discussed in terms of Baddeley's (1986) 'visuospatial sketchpad' which maintains visuospatial imagery as part of the Central Executive System.

Data were also examined for linear associations. Although a significant association was found between the MEAMS number of subtests passed "difference" scores and the DMR 'sum of social "difference" scores' ($p < .05$), this correlation was weak ($r = .5219$). It

was suggested that declining social abilities of DS clients may be weakly associated with performance on the MEAMS.

Finally, it was suggested that further longitudinal studies should be conducted that compared different forms of dementia, and particularly with vascular disorders such as multi-infarcts and stroke.

REFERENCES

Agbayewa, M.D., Weir, J., Tuokko, H. & Beattie, L. (1991), ‘Dperession in dementia: Its impact on functional ability’, *Dementia, 2:* 212-217.

Alberoni, M., Baddeley, A.D., Della Sala, S. & Logie, R. (1992), 'Keeping track of a conversation: impairments in Alzheimer's disease', *International Journal of Geriatric Psychiatry, 7:* 639-646.

Allen, P.A., Smith, A.F., Jerge, K.A. & Vires-Collins, H. (1997), 'Age differences in mental multiplication: Evidence for peripheral but not central decrements', *Journal of Gerontology*, 52; 2: P81-P90.

APA - American Psychiatric Association (1994*), ‘Diagnostic and Statistical Manual of Mental Disorders (DSM-IV). 4th ed’*, Washington DC: American Psychiatric Association, 139-143.

Atkinson, L. (1991), 'Mental retardation and WAIS-R difference scores', *Journal of Mental Deficiency Research, 35;* 537-542.

Atkinson, R.C. & Shiffrin, R.M. (1968), 'Human Memory: A Proposed System and its Control Processes' in Spence, K. & Spence, J. (eds), *The Psychology of Learning & Motivation,* New York: Academic Press.

Baddeley, A.D. (1986), *'Working Memory',* Oxford: Oxford University Press.

Baddeley, A.D. (1992), 'Working memory', *Science, 255:* 556-559.

Baddeley, A.D. (1996), *'Human Memory, Rev. ed',* Hove: Psychology Press.

Baddeley, A.D., Bressi, S., Della Sala, S., Logie, R., *et al.* (1991), 'The decline of working memory in Alzheimer's disease', *Brain, 114;* 2521-2542.

Baddeley, A.D. & Hitch, G.J. (1974), 'Working Memory' in Bower, G. (ed*), Recent Advances in Learning and Motivation. Volume III*, London: Academic Press.

Baddeley, A.D., Logie, R., Bressi, S., Della Sala, S., *et al.*. (1986), 'Dementia and working memory', *Quarterly Journal of Experimental Psychology, 38A;* 603-618.

Baddeley, A.D., Wilson, B.A. & Watts, F. (1995), *'Handbook of Memory Disorders'*, Chichester: Wiley.

Barr, O. (1990), 'Down's syndrome and Alzheimer's disease - what is the link?' *Professional Nurse, June;* 465-468.

Beck, A.T. & Steer, R.A. (1987), *'Beck Depression Inventory. Manual'*, San Antonio: Harcourt Brace Jovanovich.

Becker, J.T. (1988), 'Working memory and secondary memory deficits in Alzheimer's disease', *Journal of Clinical & Experimental Neuropsychology, 6:* 739-753.

Benton, A.L. (1977), 'Interactive effects of old age and brain disease on reaction time', *Archives of Neurology, 34;* 369-370.

Benton, A.L. & Spreen, O. (1961), 'Visual memory test; The simulation of mental incompetence', *Archives of General Psychiatry, 4;* 79-83.

Botwinick, J. (1977), 'Intellectual Abilities' in Birren, J.E. & Schaie, K.W. (eds), *Handbook of the Psychology of Aging*, New York: Van Nostrand.

Botwinick, J. (1978), *'Aging and Behavior, 2nd ed'*., New York: Springer.

Botwinick, J. & Storandt, M. (1974), *'Memory, Related Functions, and Age'*, Springfield, Illinois: C.C. Thomas.

Botwinick, J. & Storandt, M. (1980), 'Recall and recognition of old information in relation to age and sex', *Journal of Gerontology, 35;* 70-76.

Cohen, N.J. & Squire, L.R. (1981), 'Preserved learning and retention of pattern analysing skill in amnesia: dissociation of knowing how and knowing that', *Science, 210;* 207-209.

Collerton, D. (1993), 'Memory Disorders' in Greenwood, R., Barnes, M.P., McMillan, T.M. & Ward, C.D. (eds), *Neurological Rehabilitation*, Edinburgh: Churchill Livingstone.

Craik, F.I.M. (1977), 'Age Differences in Human Memory' in Birren, J.E. & Schaie, K.W. (eds), *Handbook of the Psychology of Aging*, New York: Van Nostrand.

Craik, F.I.M. (1994), 'Memory changes in normal aging', Current Directions in Psychological Science, 3; 5: 155-158.

Curran, S. & Wattis, J.P. (1989), 'Round-up: Searching for the cause of Alzheimer's disease', *Geriatric Medicine, March;* 13-14.

Daigneault, S. & Braun, C.M. (1993), 'Working memory and the self-ordered pointing task: further evidence of early prefrontal decline in normal aging', *Journal of Clinical and Experimental Neuropsychology, 15;* 6: 881-895.

Das, J.P., Davison, M., Hiscox, M., Mishra, R.K., *et al.* (1993), *'Intellectual decline and ageing: Individuals with Down syndrome compared to other individuals with mental handicaps'*, Report, Edmonton: University of Alberta.

Dickson, D.W. (2001), 'Neuropathology of Alzheimer's disease and other dementias', *Clinics in Geriatric Medicine, 17;* 2: 209-228.

Earles, J.L. & Salthouse, T.A. (1995), 'Interrelations of age, health, and speed', *Journal of Gerontology, 50;* 1: P33-P41.

Ellis, R.D., Goldberg, J.H. & Detweiler, M.C. (1996), 'Predicting age-related differences in visual information processing using two-stage queing model', *Journal of Gerontology, 51;* 3: P155-P65.

Erickson, R.C. (1978), 'Problems in the clinical assessment of memory', *Experimental Aging Research, 4:* 255-272.

Evenhuis, H.M. (1992a), *'Provisional Manual of the Dementia Questionnaire for Mentally Retarded Persons (DMR)'*, Zwammerdam: Hooge Burch.

Evenhuis, H.M. (1992b), 'Evaluation of a screening instrument for dementia in ageing mentally retarded persons', *Journal of Intellectual Disability Research, 36:* 337-347.

Eyman, R.K., Call, T.L. & White, J.F. (1991), 'Life expectancy of persons

with Down's syndrome', *American Journal of Mental Retardation, 95:* 603-612.

Fisk, A.D., Rogers, W.A., Cooper, B.P. & Gilbert, D.K. (1997), 'Automatic category search and its transfer: aging, type of search, and level of learning', *Journal of Gerontology, 52;* 2: P91-P102.

Furster, J.M. (1993), 'Frontal lobes', Current Opinions in Neurobiology, 3; 160-165.

Gath, A. (1986), 'Ageing and mental handicap', *Developmental & Child Neurology, 28:* 515-524.

Goudie, F. (1993), 'Problems of Aging' in Stokes, G. & Goudie, F. (eds*), Working with Dementia*, Oxford: Winslow Press, 29-43.

Haxby, J.V. (1989), 'Neuropsychological evaluation of adults with Down's syndrome: Patterns of selective impairment in non-demented old adults', *Journal of Mental Deficiency Research, 33:* 193-210.

Heston, L.L. (1977), 'Alzheimer's disease, trisomy 21, and myeloproliferative disorders: Associations suggesting a genetic diathesis', *Science, 196:* 322-323.

HSS - Hampshire Social Services (1989), *'Hampshire Social Services Assessment'*, Southampton: Hampshire Social Services.

Hulme, C., Lee, G. & Brown, G.D.A. (1993), 'Short-term memory

impairments in Alzheimer-type dementia: Evidence for separable impairments of articulatory rehearshal and long-term memory', *Neuropsychologia, 31:* 161-172.

Jernigan, T., Zatz, I.M. & Feinberg, K. (1980), 'Measurement of Cerebral Atrophy in the Aged' in Poon, L.W. (ed), *Aging in the 1980s.* Psychological Issues, Washington D.C.: American Psychological Corporation.

Jervis, G.A. (1948), 'Early senile dementia in mongoloid idiocy', *American Journal of Psychiatry, 105:* 102-106.

Jorm, A.F. (1990), *'The Epidemiology of Alzheimer's Disease and Related Disorders'*, London: Chapman & Hall.

Kang, J., Lemaire, H.G., Unterbeck, A., Selbaum, J.M., *et al.* (1987), 'The precursor of Alzheimer's disease amyloid A4 protein resembles a cell surface receptor', *Nature, 235:* 733-736.

Kapur, N., Thompson, S.B.N., Cook, P., Lang, D. & Brice, J. (1996), 'Anterograde but not retrograde memory loss following combined mammilliary body and medial thalamic lesions', *Neuropsychologia, 34;* 1: 1-8.

Katzman, R. (1976), 'The prevalence and malignancy of Alzheimer's disease', *Archives of Neurology, 33:* 217-8.

Kramer, N.A. & Jarvik, L. (1979), ‘Assessment of Intellectual Changes in the Elderly’ in Raskin, A. & Jarvik, L. (eds), *Psychiatric Symptoms and Cognitive Loss in the Elderly*, Washington DC,: Hemisphere.

Lamberty, G.J. & Bieliauskas, L.A. (1993), ‘Distinguishing between depression and dementia in the elderly: A review of neuropsychological findings’, *Archives of Clinical Neuropsychology, 8:* 149-170.

Levinson, A., Friedman, A. & Stamps, F. (1955), 'Variability of mongolism', *Paediatrics, 16:* 43-54.

Lezak, M.D. (1983), *‘Neuropsychological Assessment, 2nd ed’*, New York: Oxford University Press.

Maclean, W.E., Ellis, D.N., Galbreath, H.N., Halpern, L.F., *et al.*. (1991), 'Rhythmic motor behaviour of preambulatory motor impaired, Down syndrome and nondisabled children - A comparative analysis', *Journal of Abnormal Child Psychology, 19:* 319-330.

Miniszek, N.A. (1983), 'Development of Alzheimer's disease in Down's syndrome individuals', *American Journal of Mental Deficiency, 85:* 377-385.

Mitrushina, M., Uchiyama, C. & Satz, P. (1995), 'Heterogeneity of cognitive profiles in normal aging: implications for early

manifestations of Alzheimer's disease', *Journal of Clinical and Experimental Neuropsychology, 17;* 3: 374-382.

Money, E.A., Kirk, R.C. & McNaughton, N. (1992), 'Alzheimer's dementia produces a loss of discrimination but no increase in rate of memory decay in delayed matching to sample', *Neuropsychologia, 30:* 133-143.

Moody, P. & Moody, R. (1992), *'Half Left'*, Oslo: Dreyers Forlag A/S.

Morris, R.G. (1984), 'Dementia and the functioning of the articulatory loop system', *Cognitive Neuropsychology, 1:* 143-157.

Morris, R.G. (1986), 'Short-term forgetting in senile dementia of the Alzheimer's type', *Cognitive Neuropsychology, 3:* 77-97.

Morris, R.G. (1987a), 'The effect of concurrent articulation on memory span in Alzheimer-type dementia', *British Journal of Clinical Psychology, 26:* 233-234.

Morris, R.G. (1987b), 'Articulatory rehearsal in Alzheimer-type dementia', *Brain & Language, 30:* 351-362.

Morris, R.G. (1992), 'Patterns of Short-Term Memory Impairment in Alzheimer's Disease' in Backman, L. (ed), *Memory Functioning in Dementia*, Amsterdam: Elsevier, 3-25.

Morris, R.G. (1994a), 'Working memory in Alzheimer-type dementia',

Neuropsychology, 8; 4: 544-554.

Morris, R.G. (1994b), 'Recent developments in the neuropsychology of dementia', *International Review of Psychiatry, 6:* 85-107.

Norman, D.A. & Shallice, T. (1986), 'Reflections on Cognition and Parallel Distributed Processing' in McClelland, J.L. & Rumelhart, D.E. (eds), *Parallel Distributed Processing: Exploration of the Microstructure of Cognition. Volume 2*, Cambridge, Massechusetts: MIT Press, 102-110.

Nyberg, L., Backman, L., Erngrund, K., Olofsson, U. & Nilsson, L.G. (1996), 'Age differences in episodic memory, semantic memory, and priming: relationships to demographic, intellectual, and biological factors', *Journal of Gerontology, 51;* 4: P234-P40.

Palmer, C.G.S., Crank, C., Pueschel, S.M., Wisniewski, K.E., *et al.* (1992), 'Head circumference of children with Down's syndrome (0-36 months)', *American Journal of Medical Genetics, 42:* 61-67.

Parkin, A.J., Walter, B.M. & Hunkin, N.M. (1995), 'Relationships between normal aging, frontal lobe function, and memory for temporal and spatial information', *Neuropsychology, 9;* 3: 304-312.

Powell, T., Brooker, D.J.R. & Papadopolous, A. (1993), 'Test-retest reliability of the Middlesex Assessment of Mental State (MEAMS):

A preliminary investigation in people with probable dementia', *British Journal of Clinical Psychology, 32:* 224-226.

Prosser, G. (1989), 'Down's syndrome, Alzheimer's disease, and reality orientation', *Mental Handicap, 17;* 2: 50-53.

Raven, J.C., Court, J.H. & Raven, J. (1995), *'Raven Manual: Section 2. Coloured Progressive Matrices 1995 Edition'*, Oxford: Oxford Psychologists Press.

Reisberg, B. (1983), 'An Overview of Current Concepts of Alzheimer's Disease, Senile Dementia, and Age-Associated Cognitive Decline', in Reisberg, B. (ed), *Alzheimer's Disease, the Standard Reference*, The Free Press: New York.

Roper, A.H. & Williams, R.S. (1980), 'Relationship between plaques, tangles and dementia in Down's syndrome', *Neurology, 30:* 639-644.

Sahakian, B.J., Morris, R.G., Evenden, J.L., Heald, A., Levy, R., Philpot, M. & Robbins, T.W. (1988), 'A comparative study of visuospatial memory and learning in Alzheimer-type dementia and Parkinson's disease', *Brain, 111:* 698-718.

Schaie, K.W. (1958), 'Rigidity-flexibility and intelligence: a cross-sectional study of the adult life span from 20 to 70 years', *Psychological Monographs, 72;* 9: 462.

Shallice, T. (1988), *'From Neuropsychology to Mental Structure'*, New York: Cambridge University Press.

Shermann, S.L., Takesu, N., Freeman, S.B., Grantham, M., *et al.* (1991), 'Association between reduced recombination with nondisjunction', *American Journal of Human Genetics, 49:* 608-620.

Spinnler, H. & Della Sala, S. (1988), 'The role of clinical neuropsychology in the neurological diagnosis of Alzheimer's disease', *Journal of Neurology, 235:* 258-271.

Squire, L.R. (1974), 'Remote memory as affected by aging', *Neuropsychologia, 12:* 429-435.

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

St George-Hyslop, P.H., Tanzi, R.E., Polinsky, R.J., Haines, J.L., *et al.* (1987), 'The genetic defect causing familial Alzheimer's disease maps on chromosome 21', *Science, 235:* 885-890.

Stokes, G. & Holden, U. (1993), 'Dementia: Causes & Clinical Syndromes' in Stokes, G. & Goudie, F. (eds), *Working with Dementia*, Bicester: Winslow Press.

Swearer, J.M. & Kane, K.J. (1996), 'Behavioral slowing with age: boundary conditions of the generalized slowing model', *Journal of*

Gerontology, 51: 4: P189-P200.

Thompson, S.B.N. (1993), *'Eating Disorders: A Guide for Health Professionals'*, London: Chapman & Hall.

van Belle, G., Uhlmann, R.F., Hughes, J.P. & Larson. E.B. (1990), 'Reliability of estimates of change in mental status test performance in senile dementia of the Alzheimer type', *Journal of Clinical Epidemiology, 43;* 6: 589-595.

Verhaart, W.J.C. & Jelgersma, H.C. (1952), 'Early senile dementia in mongolian idiocy. Description of a case', *Folia Psychiatrica Neerlandica, 55:* 453-459.

Wechsler, D. (1981), *'Manual for the Wechsler Adult Intelligence Scale – Revised'*, New York: The Psychological Corporation.

Wechsler, D. (1988), *'Wechsler Memory Scale – Revised'*, San Antonio: Harcourt, Brace, Jovanovich.

Welford, A.T. (1977), 'Motor Performance' in Birren, J.E. & Schaie, K.W. (eds), *Handbook of the Psychology of Aging*, New York: Van Nostrand.

Wilson, B.A., Alderman, N., Burgess, P., Emslie. & Evans, J.J. (1996), *'Behavioural Assessment of the Dysexecutive Syndrome Manual'*, Bury St Edmunds: Thames Valley Test Co.

Wisniewski, K.E., French, J.H., Rosen, L.J., Koziowski, P.B., *et al.* (1983), 'Basal ganglia calcification in Down's syndrome - another manifestation of premature ageing', *Annals of the New York Academy of Sciences, 396:* 179-189.

Young, E.C. & Kramer, B.M. (1991), 'Characteristics of age-related language decline in adults with Down's syndrome', *Mental Retardation, 29;* 2: 75-79.

Zatz, L.M., Jernigan, T.L. & Ahumada, A.J.,Jr. (1982), 'White matter changes in cerebral computed tomography related to aging', *Journal of Computer Assisted Tomography, 6:* 19-23.

Zigmond, A.S. & Snaith, R.P. (1983), 'The Hospital Anxiety and Depression Scale', *Acta Psychiatrica Scandinavica, 67:* 361-370.

PART II: Evaluation of Treatment and Services (Published Papers)

Chapter 6

DESIGN AND EVALUATION OF A COMPUTERISED VERSION OF THE BENTON VISUAL RETENTION TEST

S.B.N. Thompson, E. Ennis, T. Coffin & S. Farman

Computers in Human Behavior (2007), 23, 2383-2393.

ABSTRACT

The use of computers in the administration of psychological assessments is often considered standard practice. However, the evidence clearly shows that the computerisation of test needs to be evaluated independently. The current study examined the hypothesis that a computerised administration of the Benton Visual Retention Test (BVRT) should yield comparable results to paper-and-pencil administration of this measure. Forty participants (23 females and 17 males) from a non-clinical

population were assessed using a computerised version of the BVRT and the conventional paper-and-pencil administration. Parallel forms of the test were used in the two administrations in order to eliminate practice effects. Participants found the conventional method of assessment easier to use but less fun. Importantly, performances of the participants were poorer when using the computerised version, giving rise to extreme caution hen using this method of assessment administration with a clinical population.

Available online at www.sciencedirect.com
ScienceDirect
ELSEVIER
Computers in Human Behavior 23 (2007) 2383–2393
Computers in Human Behavior
www.elsevier.com/locate/comphumbeh

Design and evaluation of a computerised version of the Benton visual retention test ☆

S.B.N. Thompson [b], E. Ennis [a,*], T. Coffin [a], S. Farman [a]

[a] *Centre for the Study of Emotion Department of Psychology, University of Portsmouth, King Henry Building, King Henry 1 Street, Portsmouth PO1 2DY, UK*
[b] *Neuropsychology Assessment and Consultancy, Portsmouth, UK*

Available online 22 May 2006

Abstract

The use of computers in the administration of psychological assessments is often considered standard practice. However, the evidence clearly shows that computerisation of each test needs to be evaluated independently. The current study examined the hypothesis that a computerised administration of the Benton visual retention test (BVRT) should yield comparable results to paper-and-pencil administration of this measure. Forty participants (23 females and 17 males) from a non-clinical population were assessed using a computerised version of the BVRT and the conventional paper-and-pencil administration. Parallel forms of the test were used in the two administrations in order to eliminate practice effects. Participants found the conventional method of assessment easier to use but less fun. Importantly, performances of the participants were poorer when using the computerised version, giving rise to extreme caution when using this method of assessment administration with a clinical population.
© 2006 Elsevier Ltd. All rights reserved.

Keywords: Attitude to computers; Benton visual retention test; Computerised assessment; Computer preferences; Memory

☆ This research was funded by the Summer Bursary Scheme, University of Portsmouth, Department of Psychology, and has been approved by the University's Ethics Committee.
* Corresponding author. Tel.: +44 23 9284 6312.
E-mail address: edel.ennis@port.ac.uk (E. Ennis).

0747-5632/$ - see front matter © 2006 Elsevier Ltd. All rights reserved.
doi:10.1016/j.chb.2006.03.014

1. Introduction

Computers have the potential to be used across a variety of settings, and for a variety of procedures, ranging from administration of psychological tests to test scoring and interpretation (Schulenberg & Yutrzenka, 2004; Weber et al., 2003). The current paper examines the potential use of computers within the administration of the Benton visual retention test (BVRT), which is often used in the clinical setting. Towards the end of the last century, computer-based psychological assessments had become almost mainstream in terms of clinicians' tools for the assessment of a range of neuropsychological conditions and sequelae. However, this has not been without comment or reservations from researchers and clinical workers alike.

Since the late 1980s, growing technological sophistication has been evidenced in the psychological assessment of patient populations (Schulenberg & Yutrzenka, 2004). In particular, this has been evident in the use of computers to assess personal sensitive information amongst students (Knapp & Kirk, 2003), administer (and interpret) the MMPI in a non-clinical sample (Pinsoneault, 1996). In addition to this, within clinical samples, computers have been successfully used to assess depression and function in primary care older patients (Kurt, Bogner, Straton, Tien, & Gallo, 2004), attention and memory in psychiatric patients (Weber et al., 2003), visual retention in neurological patients (Merten, 1999), and memory difficulties (Aharonson & Korczyn, 2004), and the use of microcomputers to assess the recovery of stroke or incompletely innervated patients (Thompson, 1987; Thompson & Coleman, 1987, 1989; Thompson, Coleman, & Yates, 1986; Thompson & Morgan, 1996).

The majority of users accept computerised psychological assessment, and agree that this holds considerable advantages over paper-and-pencil versions of assessments (e.g. speed and thus cost efficiency, consistency of administration procedures) (Weber et al., 2003). However, the implications of changes such as the removal or reduction of the human examiner from the interaction and the need for adherence to the Data Protection Acts must also be considered (Schulenberg & Yutrzenka, 2004). This has spawned guidelines for choosing suitable computerised assessments (APA, 1986; BPS, 2002; Green, Bock, Humphreys, Linn, & Reckase, 1984; JCTP, 1988; Schoenfeldt, 1989), and examinations of users' attitudes towards computers in general (Bouman, Wolters, & Wolters-Hoff, 1989).

Users' attitudes towards computers are important particularly from an ethical point of view; patients should not be exposed to procedures that they find aversive (Schulenberg & Yutrzenka, 2004; Weber et al., 2003). In addition to being important from an ethical point of view, these confounding variables such as attitudes towards computers also practically influence the process and outcome of computerised testing sessions (Schulenberg & Yutrzenka, 2004; Weber et al., 2003). Research shows that both acceptance and previous experience with computers increase user's motivation and success of the patient–computer interaction (Schulenberg & Yutrzenka, 2004; Weber et al., 2003).

Computers may be used to either generate new psychological measures, or to adapt conventional measures into computerised formats (Schulenberg & Yutrzenka, 2004). As was outlined earlier, computerised assessment is effective for a variety of purposes, within both clinical and non-clinical settings. Moreover, computerised assessment is well accepted by patients despite prior apprehension about taking the test (Weber et al., 2003), and effective even when participants had no prior computer experience (Kurt et al., 2004). How-

ever, psychometric equivalence (i.e. the applicability of the same norms, etc.) across modalities should not be assumed (Schulenberg & Yutrzenka, 2004; Williams & McCord, in press).

Effective use of computers for administration of tests has been predominantly used for verbal tests that involve reading simple questions and answering with simple responses (Williams & McCord, in press). Also, results of equivalence across modalities (computer versus paper and pencil) have been more consistent for tests in this domain (Murphy & Davidshofer, 2005; Williams & McCord, in press). The computerization of tests involving graphics and manipulation of materials on the computer screen (e.g. Ravens progressive Matrices (RPM)) has not been either as widespread or as effective (Williams & McCord, in press). For example, although methodological differences across studies must be considered, findings has been inconsistent as to whether individuals score the same on a computerised administration of Ravens progressive Matrices and a paper-and-pencil version, or whether performance is poorer on the computerised version (Williams & McCord, in press).

Overall, it is clear that the adoption of an equivalent computer-based version of an existing psychological assessment tool should be met with some caution. Needless to say, the pre-requisites of validity, reliability, specificity, consistency, etc. should be found. Then, it is necessary to make judgements about the ease of use, the comfort of use by the administrator and examinee, the relative cost of such an assessment tool, and the comparability of the scores obtained through a computerised administration of the test to those obtained through a paper-and-pencil administration. This should be evaluated for each test independently, especially if the test is visual–spatial based. This point is even more poignant in relation to the BVRT given that the test is visuo-spatial, and participants are required to actually generate (draw) their response as opposed to simply selecting a correct alternative (e.g. as is done within the Ravens Progressive Matrices).

In order to examine equivalence across modalities and some of the above issues, a computerised version of the Benton visual retention test (BVRT) (Benton, 1992) was designed. Questionnaires were also designed to evaluate key issues such as whether users find such tools easy to use or comfortable to use, or whether they prefer conventional methods of administration. Results will examine how users perform on a parallel version of the BVRT when it is administered using a computer screen and requiring responses to be input directly to the computer.

The BVRT is regarded as a reliable and robust tool in many clinical testing situations and has been used in the early detection of Alzheimer's disease (Thompson, MacDonald, & Coates, 2001), for dementia in the elderly (Jacqmin-Gadda, Fabrigoule, Commenges, Letenneur, & Dartigues, 2000), and for older neurological populations (Coman et al., 2002). Computerising such a test presents a challenge. It cannot be assumed that test characteristics such as ease of administration, reliability, and accuracy will not change. Also, given that the test is most often used within sensitive populations, it is important to examine whether participant factors such as attitudes towards computers or previous experience with computers are influential in determining whether the computerised assessment is comparable to the paper-and-pencil assessment.

The current research was inspired by Merten's (1999) work on a computer-administration of the BVRT using neurological patients. Merten's (1999) findings showed no significant differences between a group who were administered the BVRT via a computer and a group who were administered the assessment via paper and pencil. However, the fact

remains that they used a between subjects design. While these two groups were matched with regards to age, gender and aetiology, no mention is made of whether attitudes towards computers or computer experience were examined. With regard to examination of within subjects patterns, a marked decline in patients' performances was evident when the computerised version followed the conventional paper-and-pencil version (Merten, 1999). However, this may be attributable merely to fatigue effects, especially given that the sample was of a clinical nature.

The current study attempts to further clarify this research area by investigating these questions within a normal population, and overcoming the methodological issues evident within the research of Merten (1999). Specifically, a within subjects design will be used to ensure that any differences in performance are due to the effects of mode of administration (computer versus paper and pencil). Also, the method received first (computer versus paper and pencil) will be controlled in order to ensure that any differences are not simply due to fatigue effects. Also building upon the research of Merten (1999), the current study will consider the importance of users' attitudes towards computers and computer experience As was outlined earlier, both these factors should be considered ethically (Bouman et al., 1989), and they are also likely to practically influence the process and outcome of computerised testing sessions (Schulenberg & Yutrzenka, 2004; Weber et al., 2003).

Overall, the present study seeks to clarify whether findings obtained administering the BVRT via a computer are comparable to those obtained administering the test via paper and pencil. Obviously, this issue has considerable practical/clinical implications in terms of time saved via reduced examiner involvement, the financial implications of this, the benefits of computers to achieving standardisation across administrations etc. Eventually, it may be possible to develop a process whereby the BVRT may be scored by means of a computer program, however it is essential to firstly demonstrate that participant's performance on both formats is equivalent.

Although they cannot be assumed to directly translate, given the findings of Merten (1999), it is hypothesised that performance on the computerised version of the BVRT will be comparable to performance on the paper-and-pencil version of the test (H1). Secondly, given their earlier discussed importance on the success of computerised testing, performance on the computerised administration of the BVRT will be positively related to positive attitudes towards computers (H2).

2. Method

2.1. Participants

Forty individuals (23 females; 17 males) were recruited from the University's pool of volunteer participants. The age range of this non-clinical population was 19–59 years old (mean = 30.67; SD = 12.58).

2.2. Materials

A conventional paper-and-pencil version as well as a computerised version of the BVRT was performed by each participant. Firstly, the Hospital Anxiety and Depression Scale (HADS) (Snaith & Zigmond, 1983) was administered. This comprises a 14-item questionnaire used to assess the severity of both depression and anxiety in order detect

probable caseness. Cut-off points indicate whether someone is 'within the normal range' (0–7), 'mildly' (8–10), 'moderate' (11–14) or 'severely' (15–21) depressed or anxious. Psychometric properties of the instrument are satisfactory and have been well documented elsewhere (Snaith & Zigmond, 1983).

The National Adult Reading Test (NART) (Nelson, 1982) was also administered and consists of 50 words printed in two columns on a card presented to the participant. These words are relatively short in order. Although not important in the current non-clinical sample, this is important in clinical samples in that it avoids stimulus complexity adversely affecting the reading of dementing subjects. However, the words are all 'irregular' with respect to the common rules of pronunciation, to avoid participants reading by phonemic decoding rather than word recognition. The participant reads each word aloud and the number of errors made in pronunciation is recorded. There is evidence that the NART shows satisfactory internal consistency, is a valid predictor of IQ, and performance on the test is unrelated to either age or social class (Nelson, 1982). Full scale Intelligence Quotient (IQ) as assessed by the Wechsler Adult Intelligence Scale Revised (WAIS-R) can be predicted from this reading error score. The scale was therefore included in the current study to both describe the IQ of the sample given that they were recruited in a university setting, and to examine whether IQ related to participants performance on the BVRT (on either computer or paper-and-pencil administration).

The BVRT assesses visual perception, visual memory, visuo-constructive abilities, some attention difficulties and visual neglect in both clinical and research settings. The test consists of 10 designs within a stimulus booklet, with each design containing one or more (typically three) figures. Three almost equivalent forms of the BVRT are available, and four alternative methods of administration. These vary in length of exposure time, and the delay period between exposure and reproduction of the stimuli by the participant. The participant is shown each design for a set period of time, after which the design is taken away and the participant is required to reproduce the design. 'Number correct' and 'number error' scores are calculated, and compared to the normative data. Broadly speaking, the number correct is calculated in terms of how similar the participants reproduced design is to the initial design they were shown. Number of errors is calculated in terms of how much error the participant has made within their reproduced design in comparison with the initial stimuli. For the computerised administration of the BVRT within the current study, a slide show within Microsoft PowerPoint was used to present each of the designs for the specified 10 s administration. Following this, a blank screen automatically appeared and the participant was required to reproduce the designs on an A5 ActivPresenta graphics tablet (Misco, 2004) connected to the personal computer (PC). A cordless pen is used on the surface of the tablet, which communicates wirelessly with an adaptor connected to the computer's USB port. As the participant draws on the graphics tablet, a representation of their drawing appears on the computer screen. On completion of the drawing, the participant was required to click the computer mouse for the next design to appear on the screen in front of them.

Finally, participants completed the Groningen Computer Attitude Scale (GCAS) (Bouman et al., 1989) which was translated from German, a questionnaire on method preference (adapted from Merten, 1999), and a questionnaire on computer experience that was specifically constructed for this study. The GCAS is a 16-item measure of general attitude towards computers that required participants to indicate their response using a 5-point Lickert scale (i.e. ranging from 'totally agree' to 'totally disagree'). The possible score

range is 16–80, with scores above 48 considered to represent a positive attitude towards computers (Bouman et al., 1989). This instrument has satisfactory psychometric properties (Weber, Fritze, Schneider, Kühner, & Maurer, 2002). The 8-item measure of method preference asked of participants which method (paper-and-pencil versus computerised version) they could best concentrate on, which one was most pleasant, which they would prefer for further tasks of a similar nature, which one they found easier, which one they would prefer if they had to carry out such tasks for a job interview, which gave them the most fun, which they found most comfortable and finally, with which method did the time go fastest. Response options were 'paper and pencil', 'computer' or 'no difference'.

The newly constructed measure of computer experience consisted of 4 items, each of which was subdivided into two sections. Item 1 enquired whether or not participants had ever used a computer before, and if so how much experience they had of using a computer (limited, intermediate or extensive experience). Item 2 enquired whether participants used a computer regularly in their everyday life, such as for work or study, and if so, how often they used the computer (occasionally, frequently, or everyday). Item 3 asked whether participants were comfortable using computers or computerised technology, and if so, how comfortable they were (fairly, moderately or very comfortable). Item 4 asked whether participants had ever used a computerised pad to input information into a computer before, and if so, how much experience they had of using it (limited, intermediate or extensive experience). Exact answer alternatives were not given. Although there are limitations with this approach (i.e. individuals interpreting the terms differently), the aim of the questionnaire was to record the participants 'subjective' feelings regarding their experience and comfort with computers. It is participants' own feelings about their interactions with the computer that are most likely to be influential.

2.3. Procedure

The HADS was administered first to ensure that no mild levels of anxiety or depression were present within the sample. Scores above 15 (on both the anxiety and depression components of the questionnaire) indicate such affective disturbances. No participants were found in this category, and thus all proceeded to completing the NART. A code of procedure involving forms of consent and payment at each stage was strictly adhered to in this study in line with University's Ethics approval and the British Psychological Society's guidelines (BPS, 2004).

All participants subsequently performed both the computerised administration of the Benton visual retention test and the paper-and-pencil version of the test. As was noted earlier, there are four alternative methods of administering the BVRT. These all vary the length of time for which the participant is exposed to the stimuli, and the length of time between withdrawing the stimulus and the participant generating a reproduction. In the current study, administration A was adopted for both versions (i.e. the computerised and the paper and pencil). In this method of administration, stimuli were displayed for 10 s and then immediately reproduced by the participant. To eliminate order effects, participants were randomly allocated to either the BVRT or paper-and-pencil version to complete first (i.e. 'paper-and-pencil' versus 'computerised' versions).

In addition to this, Form C of the BVRT was always used within the computerised administration of the test, whilst Form D was always used within the paper-and-pencil administration. The different parallel versions of the BVRT are identical in format (see

earlier materials section for more detailed description of test format and content), but simply use different drawing designs. The use of different design drawings means that practice or memory effects across the two performances (the computer administration and the paper-and-pencil administration) are eliminated, but participants two performances can still be compared. Finally, participants completed questionnaires on computer experience, attitude to computers and method preference. These measures were completed last in order to prevent participants being too fatigued when completing the BVRT, and also to avoid the content of these questionnaires making the objectives of the study too apparent.

3. Results

Mean response scores for anxiety, depression, full IQ and the general attitudes towards computers questionnaire were 6.42 (range 1–12; SD = 2.71); 2.52 (range 0–7; SD = 1.74); 113.52 (range 102–126; SD = 4.82); and 61.6 (range 43–73; SD = 7.56), respectively. The depression, anxiety and IQ measures were included to provide information on the nature of the sample. As can be seen, participants' depression and anxiety levels were largely within a non-clinical range. IQ scores are also within a normal range. IQ scores bore no significant relation to either the number of errors or the number of correct responses made on the BVRT. Correlations ranged between −0.2 and 0.2 (all ns) regardless of the method of administration (computer versus paper and pencil). Table 1 illustrates participant's responses to the computer experience questionnaire. As can be seen from this table, the majority of individuals had some form of computer experience, used computers regularly in their everyday lives, and were comfortable using computers.

Of those who had computer experience, 10% rated it as limited, 45% rated it as intermediate, 37.5% rated it as extensive, and 7.5% neglected to indicate their level of experience. Of those who used computers regularly in their everyday lives, 2.5% rated their use of computers as occasional, 22.5% rated it as frequent, 62.5% rated it as everyday, and 12.5% neglected to rate their degree of usage. Of those who said they were comfortable using computers, 12.5% said they were fairly comfortable, 30% were moderately comfortable, and 47.5% were very comfortable and 10.0% did not say. Few individuals had experience of using computerised pads, and those who did rated their experience as either limited or intermediate.

To test possible differences in performance on the computerised versus paper-and-pencil version, a repeated measures ANOVA utilizing a 2 (type of administration: number correct within computerised administration versus number correct within paper-and-pencil administration) × 2 (order of administration: computerised first versus paper-and-pencil first) mixed design was employed. The number correct within both forms of administra-

Table 1
Percentage responses for computer experience questionnaire

	Yes	No
Have you ever used a computer before?	92.5	5.0
Do you use the computer regularly in your everyday life?	85.0	12.5
Are you comfortable using computers?	90.0	10.0
Have you ever used a computerised pad before?	17.5	82.5

tion was a within subjects factor, whilst the order of administration was a *between* subjects factor. Results showed that participants provided significantly more correct responses using the paper-and-pencil version of the test ($M = 8.35$; SE = .25) compared to the computer version of the test ($M = 6.22$; SE = .33) ($F(1,38) = 67.19$, $p < .001$, $\eta^2 = .64$). The order of administration did not exert any interactive effect ($F(1,38) = .08$; ns).

To test for differences in error responses, a second repeated measures ANOVA was employed utilizing a 2 (type of administration: number incorrect within computerised administration versus number incorrect within paper-and-pencil administration) × 2 (order of administration: computerised first versus paper-and-pencil first) mixed design was employed. The number incorrect within both forms of administration was a within subjects factor, whilst the order of administration was a *between* subjects factor. Results showed that participants provided significantly less error responses using the paper-and-pencil version of the test ($M = 1.75$; SE = .28) compared to using the computer version of the test ($M = 5.25$, SE = .57) ($F(1,38) = 57.92$, $p < .001$, $\eta^2 = .60$). Again, order of administration did not exert any interactive effect on this main effect ($F(1,38) = 0.29$; ns). Thus, results did not support our hypothesis (H1), in that the performance on the computerised version of the BVRT was significantly different to performance on the paper-and-pencil version of the test. Specifically, performance on the computer version of the test was significantly poorer than performance on the paper-and-pencil version of the test.

Contrary to Hypothesis 2, Pearson's correlation coefficient showed that the GCAS bore no significant relation to either the number of correct responses using the computer administration ($r = .15$; ns) or the number of error responses using the computer administration ($r = -.27$; ns). It should be noted that this analysis should be accepted with caution due to a lack of variability. Specifically, whilst scores on the GCAS scale within the current study ranged between 43 and 73; the majority of participants in the current study had positive attitudes towards computers. Scores on the GCAS scale can theoretically range between 16 and 80, with scores above 48 indicating positive attitudes towards computers (Weber et al., 2003).

Results indicated that computerised administration was the most 'fun' (see Table 2). However, when asked about practical factors of the preferred method, the paper-and-pencil version was easiest to concentrate on, preferred for future tasks, easier generally, and the most comfortable. Opinions appeared to be divided as to which method, if any, the time went faster with.

Table 2
Percentage responses for method preference questionnaire

	Paper and pencil	Computerised	No difference
Easiest to concentrate on	67.5	7.5	25
Most pleasant	77.5	12.5	10
Preferable for similar tasks	75	15	10
Easiest	92.5	5	2.5
Preferable for job interview tasks	80	7.5	12.5
Most fun	15	77.5	7.5
Most comfortable	77.5	2.5	20
Time went faster	45	32.5	22.5

4. Discussion

The majority of participants (70%) indicated that they were able to concentrate better on the paper-and-pencil (PAP) version of the BVRT. This was a surprising result given that computer technology is now a part of most people's lives, both in the home and work environment. Accordingly, the majority of participants in the current study indicated that they were quite comfortable using computers and had experience of using computer technology. However, attention should be drawn to the fact that this refers to 'subjective' rather than 'objective' computer experience, and different individuals may have interpreted terms such as 'comfortable' differently.

The above findings are consistent with Merten's (1999) study of Dutch speaking unselected neurological patients. Compared to the computerised version, the majority of participants rated the PAP version as more pleasant to use, easier to use, more comfortable to use, and preferable for use in future tasks of a similar nature. However, consistent with existing research in the area (Kurt et al., 2004; Weber et al., 2003), the computerised version of the conventional test received favourable evaluations in other areas. It was interesting to note that the computer version was typically considered as being more fun to use than the PAP.

In terms of test performance, despite controlling for which version was administered first, participants' performances were significantly better on the PAP than on the computer version of the same test. This poorer performance on the computerised version of the BVRT is despite participants having previous experience with computers and having positive attitudes towards computers. Additionally, although education has been shown to interact with the success of the patient–computer interaction (Weber et al., 2003), this should not have been an influential factor within the current study although it was IQ that was assessed as opposed to educational attainment. It is possible that participants find the computer version less user-friendly in terms of the display, and more confusing to use in terms of the graphics tablet and mouse. This is also consistent with Merten's (1999) study of neurological patients which is interesting since it seems that between these two studies both a non-clinical and a clinical population share performances, i.e. both populations declined when the computer version was used. Also, as the order of administration of test was randomised (i.e. computerised BVRT first versus paper-and-pencil version first), this confirms that the decline in performance on the computerised version is not simply attributable to fatigue effects.

Results of number correct and number error responses on the BVRT were examined for both versions of administration showing that the computer version yielded poorer performances. Hence, the computer version of the BVRT is less reliable than the PAP in a non-clinical population. Therefore, extreme caution should be exercised if it were to be used in a clinical population. In fact, the authors would advise against such use in its present format. This finding is in line with the opinion that performance on a computerised version of the test on visuospatial tests such as the BVRT is often significantly poorer than performance on a paper-and-pencil version of a test (Merten, 1999; Williams & McCord, in press). This reinforces the suggestion that computerisation of each test should be evaluated independently.

It may be that the method of input of participants' responses via a graphics tablet is the key to the poor performance evidenced in this study. It is suggested that responding to presented stimuli by "drawing" directly onto a special computer touch screen, as was used by Weber et al. (2003), may be better as participants are able to see in front of them their immediate response rather than via a horizontal graphics tablet. However, the cost of this sort of display is approximately double the price of the device used in this study.

It should also be noted that the format used in the current computerised version of the test is comparable to that used in the conventional paper-and-pencil administration. Specifically, with the use of the graphics tablet, the drawing tablet is placed horizontally in front of the participant. This is comparable to within the paper-and-pencil administration, where participants draw onto a piece of paper in front of them. Moreover, although a representation of the drawing appears on the computer screen in front of the participant, the participant can also see what they are drawing when they look down at the graphics tablet that they are drawing on to. Thus, this is comparable to what the participant would see when looking at their piece of paper within the paper-and-pencil administration. In future examinations, giving participants a break should be considered, particularly in view of the fact that participants found the computer version more difficult to concentrate on. Weber et al. (2003) who allowed interruptions on demand during testing, found the computerised version of the test comparable to the paper-and-pencil version of the test.

To aid scoring participants' responses, a computer program could be written to encode directly scores and hold in a database results from such testing. Although easier to administer for the examiner and more accurate to administer in terms of accurate timing of stimuli-display, it must be born in mind that impaired and perhaps older participants such as those found in a clinical population may find using such computer displays less easier to understand because of their lower computer literacy generally. It is suggested that this generation gap of computer literacy will fade as more and more people are familiar with technological advances in everyday situations. However, even in years to come, the particular characteristics and capabilities of each population will need to be borne in mind, especially when assessing within clinical situations.

Finally, it is suggested that whilst computer technology is no doubt a welcomed advance to most lifestyles, the human element should never be completely replaced in clinical situations where reassurance and the "bedside manner" provides empathy for the clinically challenged individual.

5. Conclusions

Evidence was found to suggest that participants from a non-clinical population perform less well on a computerised version of the Benton visual retention test as compared with the paper-and-pencil version. Participants rated the computer version as more fun but less easy to use. Hence, the authors concluded that the conventional administration is a more reliable and more accurate method of assessment administration.

Acknowledgement

The authors are grateful to Dr. G. Simons for her translation skills.

References

Aharonson, V., & Korczyn, A. D. (2004). Human-computer interaction in the administration and analysis of neuropsychological tests. *Computer Methods and Programs in Biomedicine, 73*, 43–53.

APA – American Psychiatric Association. (1986). *Guidelines for computer-based tests and interpretations*. Washington, DC.

Benton, S. A. (1992). Benton visual retention test manual (5th ed.). San Antonio: Harcourt Brace & Company.

Bouman, T. K., Wolters, F. J. M., & Wolters-Hoff, G. H. (1989). Een schaal om attitudes tegenover computers te meten (Computers attitude scale). *Nederands Tijdschrift voor de Psychologie, 44*, 288–292.

BPS – British Psychological Society (2002). Guidelines for the development and use of computer-based assessments. Leicester: Psychological Testing Centre.

BPS – British Psychological Society (2004). Code of conduct, ethical principles and guidelines, 9 June 2004. Leicester: British Psychological Society.

Coman, E., Moses, J. A., Jr., Chmura Kraemer, H., Friedman, L., Benton, A. L., & Yesavage, J. (2002). Interactive influences on BVRT performance level: geriatric considerations. *Archives of Clinical Neuropsychology, 17*, 595–610.

Green, B. F., Bock, R. D., Humphreys, L. G., Linn, R. B., & Reckase, M. D. (1984). Technical guidelines for assessing computerized adaptive tests. *Journal of Educational Measurement, 21*, 347–360.

Jacqmin-Gadda, H., Fabrigoule, C., Commenges, D., Letenneur, L., & Dartigues, J. F. (2000). A cognitive screening battery for dementia in the elderly. *Journal of Clinical Epidemiology, 53*, 980–987.

JCTP – Joint Committee on Testing Practices. (1988). *Code for fair testing practices in education*. Washington, DC.

Knapp, H., & Kirk, S. A. (2003). Using pencil and paper, internet and touch tone phones for self-administered surveys: does methodology matter? *Computers in Human Behavior, 19*, 117–134.

Kurt, R., Bogner, H. R., Straton, J. B., Tien, A. Y., & Gallo, J. J. (2004). Computer-assisted assessment of depression and function in older primary care patients. *Computer Methods and Programs in Biomedicine, 73*, 165–171.

Merten, T. (1999). Konventionelle und computer-getützte Durchführung von Leistungstets: der Benton-Test (Conventional and computerized test administration: The Benton test). *Zeitschrift für Differentielle und Diagnostische Psychologie, 20*, 97–115.

Misco. (2004). *ActivPresenta*, Misco Computer Products. Available from http://www.misco.co.uk.

Murphy, K. R., & Davidshofer, C. O. (2005). Psychological testing: Principles and applications. New Jersey, USA: Pearson Prentice Hall.

Nelson, H. E. (1982). *National adult reading test (NART)* (2nd ed.). *Test manual*. Berkshire, England: NFER Nelson.

Pinsoneault, T. B. (1996). Equivalency of computer-assisted and paper-and-pencil administered versions of the Minnesota Multiphasic Personality Inventory-2. *Computers in Human Behavior, 12*, 291–300.

Schoenfeldt, L. F. (1989). Guidelines for computer-based psychological tests and interpretations. *Computers in Human Behavior, 5*, 13–21.

Schulenberg, S. E., & Yutrzenka, B. A. (2004). Ethical issues in the use of computerized assessment. *Computers in Human Behavior, 20*, 477–490.

Snaith, R. P., & Zigmond, A. S. (1983). The Hospital Anxiety and Depression Scale (HADS). *Acta Psychiatrica Scandinavica, 67*, 361–370.

Thompson, S. B. N. (1987). A system for rapidly converting quadriceps contraction to a digital signal for use in microcomputer-oriented muscle therapy and stroke patient assessment schedules. *Computers in Biology & Medicine, 17*, 117–125.

Thompson, S. B. N., & Coleman, M. J. (1987). A quantitative assessment of neuromuscular function for use with unilateral cerebrovascular accident patients. *International Journal of Rehabilitation Research, 10*, 312–316.

Thompson, S. B. N., & Coleman, M. J. (1989). An interactive microcomputer-based system for the assessment and prognosis of stroke patients. *Journal of Microcomputer Applications, 12*, 33–40.

Thompson, S. B. N., Coleman, M. J., & Yates, J. (1986). Visual feedback as a prognostic tool. *Journal of Microcomputer Applications, 9*, 215–221.

Thompson, S. B. N., MacDonald, J., & Coates, T. (2001). Improving visual memory with Aricept (Donepezil Hydrochloride, E2020) in mild-to-moderate Alzheimer's disease. *Clinical Gerontologist, 24*, 55–73.

Thompson, S. B. N., & Morgan, M. (1996). *Occupational therapy for stroke rehabilitation*. London: Chapman & Hall.

Weber, B., Fritze, J., Schneider, B., Kühner, T., & Maurer, K. (2002). Bias in computerized neuropsychological assessment of depressive disorders caused by computer attitude. *Acta Psychiatrica Scandinavica, 105*, 126.

Weber, B., Schneider, B., Fritze, J., Gille, B., Hornung, S., Kühner, T., et al. (2003). Acceptance of computerized compared to paper-and-pencil assessment in psychiatric inpatients. *Computers in Human Behavior, 19*, 81–93.

Williams, J. E., & McCord, D. M. (in press). Equivalence of standard and computerized versions of the Raven Progressive Matrices Test. Computers in Human Behavior.

Chapter 7

IMPROVING VISUAL MEMORY WITH ARICEPT (DONEPEZIL HYDROCHLORIDE, E2020) IN MILD-TO-MODERATE ALZHEIMER'S DISEASE

S.B.N. Thompson, J. MacDonald & T. Coates

Clinical Gerontologist (2001), 24 (1/2), 55-73.

ABSTRACT

Sixteen patients diagnosed with mild-to-moderate Alzheimer's disease were assessed at two time points, pre- and post-treatment with Aricept, an acetylcholine esterase inhibitor. A battery of standardized neuropsychological tests was administered in addition to recording dose tolerance and maintenance. All patients significantly improved their scores on tests measuring visual memory, visuo-spatial skills, and

auditory verbal memory after an interval of approximately 16 weeks. These findings support those of past studies that found an overall increase in cognitive functioning. However, these findings are unique in finding significant improvement in specific measures of visual memory and visuo-spatial functioning.

Improving Visual Memory with Aricept (Donepezil Hydrochloride, E2020) in Mild-to-Moderate Alzheimer's Disease

Simon B. N. Thompson, BA(Hons), MPhil, PhD, PGDipIS, AFBPsS, CPsychol
John MacDonald, BSc(Hons), MSc, PhD, AFBPsS, CPsychol
Tony D. Coates, BSc(Hons), MBChB, MRCPsych

ABSTRACT. Sixteen patients diagnosed with mild-to-moderate Alzheimer's disease were assessed at two time points, pre- and post-treatment with Aricept, an acetylcholine esterase inhibitor. A battery of standardised neuropsychological tests was administered in addition to recording dose tolerance and maintenance. All patients significantly improved their scores on tests measuring visual memory, visuo-spatial skills, and auditory verbal memory after an interval of approximately 16 weeks. These findings support those of past studies that found an overall increase in cognitive functioning. However, these findings are unique in

Simon B. N. Thompson is affiliated with The Harold Kidd Unit, Chichester, England, and is Visiting Professor, Department of Psychology, University of Portsmouth, England. John MacDonald is affiliated with the Division of Psychology, School of Social Science, University of Paisley, Paisley, Scotland. Tony D. Coates is affiliated with the Harold Kidd Unit, Chichester, England.

Address all correspondence to: Professor Simon B. N. Thompson, Clinical Psychology & Neuropsychology Services, Sussex Weald & Downs NHS Trust, The Harold Kidd Unit, Blomfield Drive, Chichester, PO19 4AU, England.

A part of this paper was a poster at the British Psychological Society Centenary Annual Conference, Scottish Exhibition & Conference Centre, Glasgow, Scotland, 28-31 March 2001. The authors are grateful to M. Warren of Pfizer Pharmaceuticals UK Ltd for sponsoring the liaison nurse; also, Dr. M. Linnell, B. Meeson, J. Rosser, Dr. H. Leegood, and F. Châbaane.

© 2001 by The Haworth Press, Inc. All rights reserved.

finding significant improvement in specific measures of visual memory and visuo-spatial functioning. *[Article copies available for a fee from The Haworth Document Delivery Service: 1-800-HAWORTH. E-mail address: <getinfo@haworthpressinc.com> Website: <http://www.HaworthPress.com> © 2001 by The Haworth Press, Inc. All rights reserved.]*

KEYWORDS. Alzheimer's disease, Aricept, dementia, donepezil hydrochloride, memory performance, visual memory

INTRODUCTION

Dementia is a progressive illness characterised by cognitive decline, behavioural and psychological disturbances, and deterioration of occupational or social functioning (Thompson, 1997). The World Health Organisation (1992) defines dementia as 'a syndrome due to disease of the brain, usually of a chronic and progressive nature, in which there is impairment of multiple higher cortical functions.' Progression is particularly evident in Alzheimer's disease (AD) as increasing impairment of memory storage and retrieval develops gradually into a global impairment of cognition (Mohs & Ferris, 1998; Birks & Melzer, 1999).

Among the many neurological and neurochemical abnormalities that develop in AD are cell loss and malfunctioning of cholinergic neurones. Acetylcholine is an important neurotransmitter associated with memory. Alterations in the enzyme systems involved in cholinergic function ie choline acetyl transferase (Chat) and acetyl choline esterase (AchE) together with loss of cholinergic neurons are amongst the most consistent findings in Alzheimer's disease (Sutherland et al., 1995).

Aricept, an acetylcholinesterase inhibitor, acts to increase the availability of acetylcholine within the synapses of neurons which remain intact in the brains of individuals suffering from Alzheimer's disease (Weinstock, 1999). This offers the possibility of improved neuronal function.

Tacrine was the first compound approved in the United States and worked in this way. However, it caused severe side effects (Conway, 1998; Qizilbash et al., 1998). Other drugs followed and have been variously evaluated; for example, metrifonate (Becker et al., 1998; Cummings, 1998; Cummings et al., 1998; McKeith, 1998; Kaufer, 1998; Tariot, 1998); piracetam (Deberdt, 1994; Gouliaev, 1994; Israel et al., 1994; Meilke, 1996; Mondadori, 1996; Zirm et al., 1994); selegiline (Freedman et al., 1998; Lawlor et al., 1997; Sano et al., 1997); and velnacrine maleate (Antuono, 1995).

The effects of medication on visual memory deficits and visuo-spatial impairment in Alzheimer's disease have not been widely documented. Reports have tended to focus on identification deficits (Dixon et al., 1999) rather than deficits in visual memory, per se, or have not considered drug regimes (Eslinger et al., 1988). In particular, the pathological correlates of visual agnosia have been investigated in Alzheimer's disease. Giannakopoulos and colleagues (1999) have demonstrated that senile plaques do not represent a pathological correlate of visual agnosia. This suggests that visual abnormalities are perhaps an early feature of Alzheimer's disease, or alternatively, that senile plaque development is a secondary phenomenon in terms of the pathophysiology of Alzheimer's disease. This is a view that has gained further recent support from workers at University of Southern California and Northwestern University, Illinois. They propose that Alzheimer's disease may be related to the formation of toxic proteins rather than the build up of plaques (Klein et al., 2001).

Similarly, there appears to be no literature on auditory verbal memory deficits (except in the general population during learning exercises-Crockett et al., 1992) but not in Alzheimer's disease, apart from inclusion as part of a global cognitive test battery (Rogers et al., 1998). It is believed that this study is unique in examining the effects of an anti-dementia drug on both visual memory deficits and visuo-spatial skills, and on auditory verbal memory functioning.

EXAMINING THE EFFECTIVENESS OF ARICEPT

One of the newer drugs for the treatment of AD is Aricept (Donepezil Hydrochloride, E2020). It is argued that donepezil hydrochloride has been shown to have modest benefits though with small but significant improvements in cognitive functioning with 5 or 10 mg per day doses (Bryson & Benfield, 1997). However, some authors are more cautious and even suggest that there have been unfounded claims of improvement in such patients (Gray, 1997). Others report several side-effects of its use, such as vomiting and diarrhoea during the titration period (Delagarza, 1998), and Aricept-induced nightmares (Barner & Gray, 1998). Whilst some benefits of its prescription to those with AD have been acknowledged, all benefit seems to be lost when it is discontinued; although this does seem to be a common feature of most of the preparations for AD use (Delagarza, 1998).

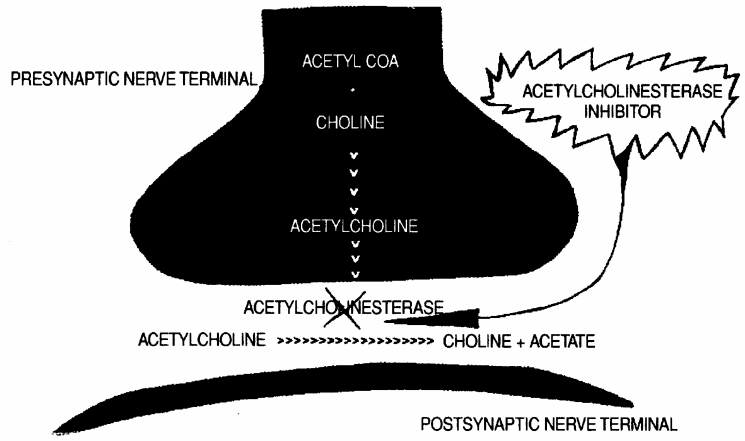
The results of three trials suggest a small beneficial effect of donepezil hydrochloride in improving cognitive function: at a 5 mg per day dose, improvements were evidenced in the mid-range of the 70-point Alzheimer's Disease Assessment Scale-Cognitive Scale (ADAS-cog) (Birks & Melzer, 1999). The authors report the results of two further trials that show some improvement in global clinical state (assessed by an independent clinician) in those treated with donepezil hydrochloride compared to placebo. The patient's own rating of their Quality of Life showed no benefit of donepezil hydrochloride compared with placebo. There were significantly more withdrawals before the end of treatment from the 10 mg per day (but not the 5 mg per day) donepezil hydrochloride group compared with placebo, which may have resulted in some over-estimation of beneficial changes at 10 mg per day in progressively declining characteristics, as last available measures were used in analysis.

Barner and Gray (1998) concluded that donepezil hydrochloride is an effective symptomatic treatment for some patients with mild-to-moderate AD and appears to be a safe alternative to tacrine, given its convenient once-daily dosing, minimal adverse effects, and lower total cost.

Rogers and Friedhoff (1996; 1998) have shown improvements in ADAS-cog scores using donepezil hydrochloride and report good tolerance of patients with no evidence of hepatotoxicity. In a further investigation by Rogers and Friedhoff (1998), a three-phase study sought to examine the efficacy and safety of using donepezil hydrochloride in the treatment of patients with mild to moderately severe AD. The authors considered the relationships between plasma donepezil hydrochloride concentrations, inhibition of red blood cell acetylcholinesterase activity, and clinical response. This was a 12-week, double-blind, placebo-controlled, parallel-group trial with a single-blind washout. Out-patients at 23 centres in the United States were randomized to receive placebo, 5 mg of donepezil hydrochloride, or 10 mg of donepezil hydrochloride (5 mg per day during week 1 then 10 mg per day thereafter) administered once daily at bedtime. Primary efficacy of donepezil hydrochloride was measured using the ADAS-cog scale and Clinician's Interview-Based Impression of Change (CIBICplus) including caregiver information. (See Figure 1.)

A total of 468 patients entered the study, more than 97% of who were included in the intention-to-treat (end point) analyses. The authors state that the use of donepezil hydrochloride produced statistically significant improvements in ADAS-cog, CIBICplus, and Mini-Mental State

FIGURE 1. Action of Acetylcholinesterase Inhibitor



Examination scores, relative to placebo. There was a statistically significant positive correlation between plasma concentrations of donepezil hydrochloride and acetylcholinesterase inhibition. A plateau of inhibition (80%-90%) was reached at plasma donepezil hydrochloride concentrations of 50 ng/mL. The correlation between plasma drug concentrations and both ADAS-cog ($p < .001$) and CIBICplus ($p = .006$) were also statistically significant, as was the correlation between red blood cell acetylcholinesterase inhibition and change in ADAS-cog ($p < .001$) and CIBICplus ($p = .005$). They concluded that donepezil hydrochloride (5 and 10 mg) administered once daily, is a well-tolerated and efficacious agent for treating patients with mild to moderately severe symptoms of Alzheimer's disease (Rogers et al., 1998).

Similar claims have been made for another acetylcholinesterase inhibitor, ENA 713 (Rivastigmine Tartrate). Corey-Bloom and colleagues (1998), for the ENA 713 B352 Study Group (supported by Novartis Pharmaceuticals Corporation), stated that on the ADAS-cog, high-dose ENA 713 produced the largest drug-*vs*-placebo difference that has been reported to date (m/s received 08 May 1998) for a dementia drug (4.94 points). Some gastrointestinal adverse events were noted but these were apparently self-limited and of mild to moderate intensity.

NEW STUDY

At the end of 1998, Pfizer Pharmaceuticals, the manufacturers of Aricept, agreed to donate funds to Sussex Weald & Downs National Health Service Trust (SWD) in order to recruit a liaison nurse to support the prescription of Aricept to selected patients, and also towards evaluating assessment tools and psychosocial impact of Aricept. This part of the study (manuscript in preparation) aimed to provide:

- Anonymised outcome data on the progress of all cases treated with Aricept;
- Exploration of monitoring tools, functional ability and carer satisfaction;
- Identification of best practice in the management of mild-to-moderate AD.

In addition, SWD employed a Consultant Clinical Neuropsychologist with specific expertise in the field of visual memory deficit, who investigated visual memory deficits in relation to Alzheimer's disease and the effects of Aricept. Key questions for this part of the study were:

- Do patients with mild to moderate AD have visual memory deficits?
- Does Aricept have an effect on the progress of visual memory deficits?

METHOD

Collecting quantifiable data, a repeated measures design (pre- and post-intervention testing) was implemented. This allowed patients in the trial to act as their own controls. The WSHA (1999) guidelines outlined the criteria for prescribing Aricept, which at that time, were constrained by the drug budget. Potentially, this would have allowed for a comparison group of patients to be used in this study, i.e., those patients waiting to be prescribed but not yet on Aricept. However, the NICE (2001) guidelines have enabled Aricept to be more widely available with the consequence of no longer having ethical reasons for holding a waiting group of patients. Inevitably, important data about the rate of change of untreated patients with Alzheimer's disease over the 16-week period have not been possible to collect.

Procedure

Data was collected from neuropsychological testing and clinical interview of patients who were about to be prescribed Aricept and then repeated after they had taken Aricept approximately 16 weeks later. Initial dosage was 5 mg (mean days on 5 mg = 84.9) followed by 10 mg when tolerated (mean days on 10 mg = 24.9).

Sample

The sample consisted of 16 patients (6 male, 10 female), ranging in age from 60 to 85 years of age, with an average age of 72.5 years (sd = 6.69). Of the 16 patients, 3 (18.75%) were aged 60-69 years, 12 (75%) were aged 70-79 years, and 1 (6.25%) was aged 85 years. They were recruited from General Practitioners' referrals to Old Age Psychiatrists within the SWD region. Patients' full scale IQs ranged from 56-105 (mean = 78.3; sd = 12.52) as assessed by the Wechsler Adult Intelligence Scale Revised (WAIS-R) (Wechsler, 1981) (Table 1). Estimates of patients' premorbid IQ were assessed using the National Adult Reading Test (NART) (Nelson, 1992), range 77-117 (mean = 96.69; sd = 11.22) (Table 1). These were found, on average to be in the abnormality range (ie discrepancy between estimated premorbid IQ and current IQ compared with "normals") of 11.3%. All patients satisfied the following inclusion criteria:

- Living outside hospital and likely to remain so for at least a year;
- Diagnosis of AD using DSM IV or ICD 10 classification;
- Mini-Mental State Examination score of 10-26;
- Aged 50 years old or over;
- Living with a carer or visited by a carer at least weekly.

Criteria for exclusion from the study were:

- Dementia due to other causes, e.g., vascular dementia, Cerebrovascular Accident, Down's syndrome, Creutzfeld Jakob's disease;
- Multiple Sclerosis or other neuro-degenerative disease;
- Living alone without visiting family or carers;
- On drugs with significant anti-cholinergic effects;
- Primary diagnosis of depression.

TABLE 1. Raw Test Scores for Current and Premorbid Intelligence Measures

PATIENT	AGE	WAIS-R			NART		
		FIQ	VIQ	PIQ	FIQ	VIQ	PIQ
01 m	74	070	073	070	092	092	094
02 f	69	069	074	064	090	090	091
03 m	71	082	087	075	085	085	087
04 m	78	071	067	078	087	087	089
05 f	85	075	078	073	095	094	096
06 m	60	080	059	069	077	078	080
07 f	77	081	082	078	117	115	116
08 f	73	088	082	105	091	091	093
09 f	78	056	058	062	113	111	112
10 f	67	076	072	084	097	097	098
11 f	72	058	058	065	108	107	108
12 m	78	105	104	105	092	092	094
13 m	74	086	085	092	098	098	099
14 f	68	082	103	088	095	094	096
15 f	76	078	082	073	095	094	095
16 f	79	096	095	099	115	113	114

KEY: WAIS-R = Wechsler Adult Intelligence Scale (Revised)
FIQ = Full Scale Intelligence Quotient
VIQ = Verbal Intelligence Quotient
PIQ = Performance Intelligence Quotient
NART = National Adult Reading Test

Table 2 shows the means and standard deviations of the NART and WAIS-R scores. The first analysis conducted was to test whether there was a difference in the premorbid estimate of IQ and the current IQ level. Using a series of paired 't' tests it was found that for all measures, current IQ levels were significantly lower than the premorbid scores. This confirms the Alzheimer's diagnosis and the impact on cognitive functioning.

Outcome Measures

Data collection was designed to limit, as far as possible, the amount of time spent assessing the client (to avoid fatigue in the patient) and taking up the carer's time.

TABLE 2. Comparison of the Premorbid (NART) and Current (WAIS-R) IQs

	NART Mean (sd)	WAIS-R Mean (sd)	t	Df	P
Full IQ	96.69 (11.22)	78.31(12.52)	4.099	15	< 0.001
Verbal IQ	96.13 (10.43)	78.69 (14.42)	3.966	15	< 0.01
Performance IQ	97.63 (10.07)	80.0 (14.12)	4.151	15	< 0.01

Testing for Cognitive Functioning (45 mins)

- National Adult Reading Test for estimating premorbid IQ (Nelson, 1992);
- Wechsler Adult Intelligence Scale-Revised subtests: digit span, arithmetic, similarities, picture completion, picture arrangement, block design, object assembly) for a cognitive functioning profile and overall IQ score (Wechsler, 1981);
- Hospital Anxiety and Depression Scale (HADS) (Zigmond & Snaith, 1983) for assessing levels of anxiety and depression.

Testing for Neuropsychological Functioning (45 mins)

- Benton Visual Retention Test (BVRT) (Benton Sivan, 1992) for assessing visual memory and visuo-spatial skills;
- Wechsler Memory Scale-Revised (WMS-R) (Auditory Verbal Memory subtest) (Wechsler, 1988) for assessing auditory verbal memory functioning.

The WAIS-R and NART were administered in preference to the newer WAIS-III and NART-R since no reliable and validated norms were available at the time for comparing current IQ with estimates of premorbid IQ using these newer measures. In contrast, reliable comparisons are possible using the former tools. With respect to discrepancies between WAIS-R and NART IQ scores, there have been several reports on demented and control subjects (Hart et al., 1986; Stebbins et al., 1990a).

Particularly with moderately to severely demented subjects, such as those participating in this study, Stebbins and colleagues (1990b) have

suggested that NART scores are influenced by dementia, even in those with only mild dementia; and if there are accompanying language deficits (Stebbins et al., 1990a), the NART appears to underestimate IQ. Hence, a large discrepancy between current IQ (as tested by the WAIS-R) and estimated premorbid IQ (as tested by the NART) might indicate a low level of intellectual functioning as associated commonly with dementia. In fact, the low NART IQ estimate would tend to mask the *true* discrepancy and therefore, the *greater* degree of lower functioning.

Several other authors have reported NART performances to be relatively resistant to dementia of the Alzheimer's type (Nebes et al., 1984; O'Carroll et al., 1987; O'Carroll & Gilleard, 1986), and to a variety of other organic conditions, eg alcoholic dementia, multi-infarct dementia, and closed-head injury (Crawford et al., 1988).

RESULTS

Before analysing the changes in visual memory scores the patients' pre-test scores on the BVRT were compared with the norms as published. All patients scored below the norm for their age and premorbid IQ level (Benton Sivan, 1992), on the number correct and the number of error measures. In order to test the extent to which there were any changes in the various measures of visual memory functioning a series of paired 't' test were conducted comparing the BVRT scores prior to treatment with the drug, and the measures taken 16 weeks into treatment. Tables 3 and 4 show the raw scores, and Table 5 shows the means and standard deviations and the outcomes of these tests. There was significant improvement over the treatment period for both the number of correct responses (BVRT-C) ($p < 0.05$) and for the size measure (BVRT-size) ($p < 0.05$) (Figure 2). There were no significant changes in the other measures of the BVRT.

In addition there was evidence of significant improvement in auditory verbal memory functioning as tested by the WMS–R. Both immediate ($p < 0.001$) and delayed ($p < 0.002$) recall showed significant improvement over the 16 week period of the trial (Figure 3). Table 6 shows the pre- and post-treatment means and standard deviations.

Although these changes in visual and auditory memory performance were observed, there were no significant changes on the anxiety and depression measures of the HADS (Table 7). Hence the changes in memory functioning cannot be attributed to a general improvement in mood.

TABLE 3. Raw Test Scores for Assessments 1 and 2

PATIENT	BVRT-1		BVRT-2		HADS-1		HADS-2		WMS-1		WMS-2	
	Corr	Err	Corr	Err	Anx	Dep	Anx	Dep	IR	DR	IR	DR
01	00	20	00	20	08	06	02	04	00	00	03	00
02	00	29	01	21	06	05	04	04	01	01	09	05
03	00	30	01	25	13	11	12	17	05	02	11	10
04	01	21	02	19	08	09	07	01	03	03	05	04
05	01	20	01	22	00	01	02	03	02	00	04	01
06	01	26	01	27	07	04	04	03	05	00	07	00
07	01	20	02	22	04	03	05	00	05	03	10	06
08	01	12	03	11	02	07	04	01	06	04	09	07
09	00	22	01	21	04	03	02	02	03	00	01	03
10	02	17	01	24	12	06	15	07	03	00	03	03
11	00	24	00	22	03	00	04	05	01	00	03	02
12	03	12	04	09	09	07	11	09	22	18	26	26
13	01	19	02	18	03	02	03	00	05	01	07	01
14	03	14	02	10	06	00	08	00	07	00	14	01
15	02	20	02	19	06	01	09	04	00	00	03	02
16	04	15	04	12	14	11	12	09	05	00	07	00

KEY: BVRT-1 (2) = Benton Visual Retention Test –Assessment 1 (2)
Corr = Number of Correct Responses
Err = Error Score
HADS-1 (2) = Hospital Anxiety and Depression Scale–Assessment 1 (2)
Anx = Anxiety Score
Dep = Depression Score
WMS-1 (2) = Wechsler Memory Scale (Revised)–Assessment 1 (2)
IR = Immediate Recall Score
DR = Delayed Recall Score

DISCUSSION

Findings were extremely encouraging and are consistent with past indicators of improved cognitive functioning following Aricept prescription. It is believed that these findings are unique in showing specific improvements in: (i) visual memory and visuo-spatial skills, and (ii) auditory verbal memory functioning.

The changes in visual memory functioning were that the patients improved in both their overall score and in their reproduction of the correct sizes of the test stimuli. A question that this raises is whether this indicates a global effect of the treatment on visual memory performance or

TABLE 4. Raw Component Scores for the Benton Visual Retention Test

Pt	BVRT-1						BVRT-2					
	Mis	Rot	Per	Om/Ad	Dis	Siz	Mis	Rot	Per	Om/Ad	Dis	Siz
01	00	00	00	15	05	00	00	00	00	13	07	00
02	07	00	00	03	16	03	04	01	00	07	09	00
03	03	00	00	11	15	01	00	00	00	14	11	00
04	00	01	00	10	10	00	00	00	00	06	13	00
05	01	01	00	13	05	00	00	01	00	11	10	00
06	03	00	00	05	15	03	04	01	00	10	10	03
07	00	00	00	05	13	02	01	00	00	10	11	00
08	03	00	00	01	08	00	03	01	00	01	06	00
09	01	00	00	07	13	01	00	00	00	13	08	00
10	01	01	00	04	11	00	06	00	00	04	14	00
11	01	00	00	14	08	01	01	00	00	09	12	00
12	04	04	00	00	04	00	02	01	00	01	05	00
13	01	00	00	13	05	00	00	01	00	07	10	00
14	00	04	00	05	05	00	00	01	00	05	04	00
15	03	00	00	09	08	00	00	02	00	12	05	00
16	02	02	00	05	05	01	01	00	00	04	06	01

KEY: BVRT-1 (2) = Benton Visual retention Test– Assessment 1 (2)
Mis = Misplacement
Rot = Rotation
Per = Perseveration
Om/Ad = Omission/Addition
Dis = Distortion
Siz = Size

whether the treatment affects specific aspects of visual memory. The indications from the other visual memory data are that they impact on global performance. Although none of the other measures from the BVRT improved significantly, as can be seen in Table 2 almost all measures changed in a positive direction.

Patients significantly improved on both the immediate and delayed recall of the WMS-R, indicating improvement in auditory verbal memory. This supports past reports that show improvement in overall memory functioning (Corey-Bloom et al., 1998). Hence to answer the original research questions, we can state that clients with mild to moderate AD do suffer from visual memory deficits, but that these are amenable to treatment with Aricept. The treatment also has a positive impact on other areas of memory. However, a number of questions remain to be

TABLE 5. Comparison of the Pre- and Post-Test Measures of the Benton Visual Retention Test

	Pre-Mean (sd) Median	Post-Mean (sd) Median	t	df	P	% Patients (Sign Test)
BVRT-C	1.25 (1.24) 1.00	1.69 (1.20) 1.00	−2.15	15	< 0.05	87.5 (X = 2; T < = 2; p < .05)
BVRT-E	20.06 (5.40) 20.00	18.88 (5.51) 20.00	1.41	15	.179 (ns)	68.8 (X = 4; T < = 3; ns)
BVRT-mis	1.88 (1.89) 1.00	1.38 (1.89) 0.00	1.02	15	.325 (ns)	50.0 (X = 3; T < = 2; ns)
BVRT-rot	0.81 (1.38) 0.00	0.56 (0.63) 0.00	.696	15	.497 (ns)	31.3 (X = 5; T < = 1; ns)
BVRT-om	7.5 (4.73) 5.00	7.94 (4.20) 7.00	−.475	15	.642 (ns)	37.5 (X = 6; T < = 3; ns)
BVRT-dis	9.13 (4.21) 8.00	8.81 (3.06) 9.00	.325	15	.749 (ns)	50.0 (X = 8; T < = 4; ns)
BVRT-size	0.75 (1.06) 0.00	0.25 (0.77) 0.00	2.24	15	< 0.05	100.0 (X = 0; T < = 0; p < .05)

KEY:
BVRT-C - the number of correct responses
BVRT-E - the number of error responses
BVRT-mis - the number of misplacement errors
BVRT-rot - the number of rotation errors
BVRT-per - the number of preservation errors
BVRT-om - the number of omission or addition errors
BVRT-dis - the number of distortion errors
BVRT-size - the number of size errors
% Patients - percent of patients with improved/stable score changes

answered. For example, (i) are these early gains sustained in the longer term? and (ii) are the effects on visual memory, specific or general?

New evidence (Inglis, 2001) suggests that sometimes improvement in cognitive functioning may be initially delayed and that switching to alternative agents can be beneficial. Any new drug that promises to improve the devastating effects of AD are cautiously welcomed. How-

FIGURE 2. Visual Memory Improvements (BVRT)

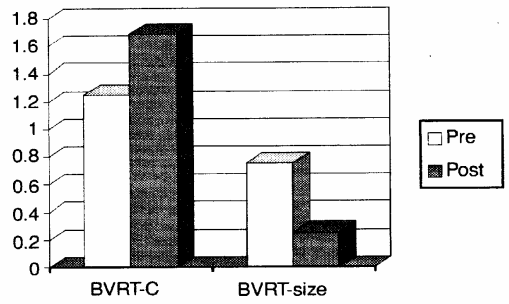
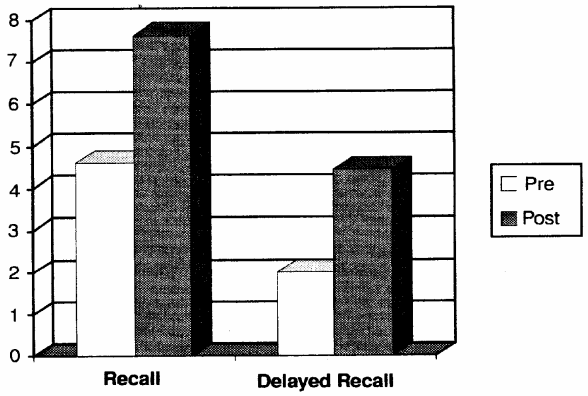


FIGURE 3. Auditory Verbal Memory Improvements (WMS-R)



ever, it is difficult to discern whether or not often subtle improvements in a patient's cognitive functioning is due solely to the drug (e.g., Skurla et al., 1998). Differential diagnoses (particularly between depression and dementia) make the process difficult; licensing of drugs is also another process that has to be properly addressed, and lastly, ethical approval. It is hoped that this new generation of ACI drugs will be much more promising than previous crusades.

TABLE 6. Comparison of the Pre- and Post-Test Measures from the Wechsler Memory Scale

	Pre-Mean (sd) Median	Post-Mean (sd) Median	t	df	P	% Patients (Sign Test)
Immediate Recall	4.56 (5.12) 3.00	7.63 (6.05) 7.00	−4.87	15	< 0.001	93.8 (X = 1; T < = 2; p < .01)
Delayed Recall	2.00 (4.47) 0.00	4.44 (6.41) 2.00	−3.86	15	< 0.002	100.0 (X = 0; T < = 1; p < .01)

KEY: % Patients = Percent of patients with improved/stable score changes

TABLE 7. Comparison of the Pre- and Post-Measures of the Hospital Anxiety and Depression Scale

	Pre-Mean (sd) Median	Post-Mean (sd) Median	t	df	P	% Patients (Sign Test)
Anxiety	6.56 (3.99) 6.00	6.50 (4.18) 4.00	.099	15	.922(ns)	43.8 (X = 7; T < = 3; ns)
Depression	4.75 (3.62) 6.00	4.31 (4.47) 3.00	.479	15	.639(ns)	56.3 (X = 6; T < = 3; ns)

KEY: % Patients = Percent of patients with improved/stable score changes

CONCLUSIONS

Sixteen patients who were prescribed Aricept showed significant improvement ($p < .05$) on measures of visual memory and visuo-spatial functioning, as measured by the BVRT. Significant evidence was found for improvement on the immediate recall ($p < .001$) and the delayed recall ($p < .002$) of the auditory verbal memory functioning, as tested by the WMS-R. Levels of anxiety and depression (measured by the HADS) were unchanged. These findings supported those of past studies that found an overall increase in cognitive functioning; however, in addition they identify, notably, areas of auditory verbal and visual memory, as specific areas of improvement.

REFERENCES

Antuono PG (1995) Effectiveness and safety of velnacrine for the treatment of Alzheimer's disease. A double-blind, placebo-controlled study. Mentane Study Group. *Archives of Internal Medicine* 155; 16: 1766-1772.

Barner EL & Gray SL (1998) Donepezil use in Alzheimer's disease. *Annals of Pharmacotherapy* 32; 1: 70-77.

Becker RE, Colliver JA, Markwell SJ, Moriearty PL, Unni LK & Vicari S (1998) Effects of metrifonate on cognitive decline in Alzheimer's disease: A double-blind, placebo-controlled, 6-month study. *Alzheimer Disease & Associated Disorders* 12 (1): 54-57.

Benton Sivan SA (1992) *Benton Visual Retention Test*. San Antonio: The Psychological Corporation.

Birks JS & Melzer D (1999) Donepezil for mild and moderate Alzheimer's disease. *The Cochrane Library* 1: 1-12.

Bryson HM & Benfield P (1997) Donepezil. *Drugs & Aging* 10; 3: 234-241.

Conway EL (1998) A review of the randomized controlled trials of tracrine in the treatment of Alzheimer's disease: Methodologic considerations. *Clinical Neuropharmacology* 21 (1): 8-17.

Corey-Bloom J, Anand R & Veach J (1998) A randomized trial evaluating the efficacy and safety of ENA 713 (rivastigmine tartrate), a new acetylcholinesterase inhibitor, in patients with mild to moderately severe Alzheimer's disease. *International Journal of Geriatric Psychopharmacology* 1: 55-65.

Crawford JR, Besson JAO & Parker DM (1988) Estimation of premorbid intelligence in organic conditions. *British Journal of Psychiatry* 153: 178-181.

Crockett DJ, Hadjistavropoulos T & Hurwitz T (1992) Primary and recency effects in the assessment of memory using the Rey Auditory Verbal Learning Test. *Archives of Clinical Neuropsychology* 7: 97-107.

Cummings JL (1998) Metrifonate: Overview of safety and efficacy. *Pharmacotherapy* 18 (2 Pt 2): 43-46.

Cummings JL, Cyrus PA, Bieber F, Mas J, Orazem J & Gulanski B (1998) Metrifonate treatment of the cognitive deficits of Alzheimer's disease. Metrifonate Study Group. *Neurology* 50 (5): 1214-1221.

Deberdt W (1994) Interaction between psychological and pharmacological treatment in cognitive impairment. *Life Science* 55:2057-2066.

Delagarza VW (1998) New drugs for Alzheimer's disease. *American Family Physician* 58; 5: 1175-1182.

Dixon MJ, Bub DN, Chertkow H & Arguin M (1999) Object identification deficits in dementia of the Alzheimer type: Combined effects of semantic and visual proximity. *Journal of the International Neuropsychological Society* 5: 330–345.

Eslinger PJ, Pepin L & Benton AL (1988) Different patterns of visual memory error occur with aging and dementia. *Journal of Clinical & Experimental Neuropsychology* 10: 60-61.

Freedman M, Rewilak D, Xerri T, Cohen S, Gordon AS, Shandling M & Logan AG (1998) L-deprenyl in Alzheimer's disease. Cognitive and behavioural effects. *Neurology* 50: 660-668.

Giannakopoulos P, Gold G, Duc M, Michel J-P, Hof PR & Bouras C (1999) Neuroanatomic correlates of visual agnosia in Alzheimer's disease. A clinicopathologic study. *Neurology* 52: 71-77.

Gouliaev AH & Senning A (1994) Piracetam and other structurally related nootropics. *Brain Research Reviews* 19 (2): 180-222.

Gray S (1997) Advertisements for Donepezil (Aricept) in the BMJ. Advertisement suggests an unrealistic improvement in mental status. *British Medical Journal* 314; 7093: 1555.

Hart S, Smith CM & Smith M (1986) Assessing intellectual deterioration. *British Journal of Clinical Psychology* 25: 119-124.

Inglis (2001) Switching acetylcholinesterase inhibitor therapy in AD. *Progress in Neurology & Psychiatry* 5; 1: 24-25.

Israel L, Melac M, Milinkevitch & Bubos G (1994) Drug therapy and memory training programs: A double-blind randomized trial of general practice patients with age-associated memory impairment. *International 6: Psychogeriatrics* 155-170.

Kaufer D (1998) Beyond the cholinergic hypothesis: The effect of metrifonate and other cholinesterase inhibitors on neuropsychiatric symptoms in Alzheimer's disease. *Dementia & Geriatric Cognitive Disorders* 9; Suppl 2: 8-14.

Klein WL, Krafft GA & Finch CE (2001) Targeting small Abeta oligomers: The solution to an Alzheimer's disease conundrum? *Trends in Neurosciences* 24; 4: 219-224.

Lawlor BA, Aisen PS, Green C, Fine E & Schneider J (1997) Selegiline in the treatment of behavioural disturbance in Alzheimer's disease. *International Journal of Geriatric Psychiatry* 12: 319-322.

McKeith IG (1998) The clinical trial protocol of Metrifonate in Alzheimer's Trial (MALT). *Dementia & Geriatric Cognitive Disorders* 9; Suppl 2: 2-7.

Meilke R (1996) Neuroprotektive Therapie des Morbus Alzheimer? [Neuroprotective therapy of Alzheimer's disease?] *Deutsche Medizinische Wochenschrift* 121 (48): 1515.

Mohs RC & Ferris SH (1998) Measuring response to treatment in Alzheimer's disease: What constitutes meaningful change? *International Journal of Geriatric Psychopharmacology* 1: Suppl 1: S7-S14.

Mondadori C (1996) Nootropics: Preclinical results in the light of clinical effects; comparison with tacrine. *Critical Reviews in Neurobiology* 10 (3-4): 357-370.

Nebes RD, Martin DC & Horn LC (1984) Sparing of semantic memory in Alzheimer's disease. *Journal of Abnormal Psychology* 93: 321-330.

Nelson HE (1992) *National Adult Treading Test Manual. 2nd ed.* Oxford: NFER-NELSON.

NICE-National Institute for Clinical Excellence (2001) *Guidance on the Use of Donepezil, Rivastigmine and Galantamine for the Treatment of Alzheimer's Disease.* London: NICE.

O'Carroll RE, Bailie EM & Whittick JE (1987) Does the National Adult Reading Test hold in dementia? *British Journal of Clinical Psychology* 26: 315-316.

O'Carroll RE & Gilleard CJ (1986) Estimation of premorbid intelligence in dementia. *British Journal of Clinical Psychology* 25: 157-158.

Qizilbash N, Whitehead A, Higgins J, Wilcock G, Schneider L & Farlow M (1998) Cholinesterase inhibition for Alzheimer disease: A meta-analysis of the tacrine tri-

als. Dementia Trialists' Collaboration. *Journal of the American Medical Association* 280 (20): 1777-1782.

Rogers SL, Doody RS, Mohs RC & Friedhoff LT (1998) Donepezil improves cognition and global function in Alzheimer's disease: A 15-week, double-blind, placebo-controlled study. Donepezil Study Group. *Archives of Internal Medicine* 158; 9: 1021-1031.

Rogers SL & Friedhoff LT (1996) The efficacy and safety of donepezil in patients with Alzheimer's disease: Results of a US multicentre, randomized, double-blind, placebo-controlled trial. The Donepezil Study Group. *Dementia* 7; 6: 293-303.

Rogers SL & Friedhoff LT (1998) Long-term efficacy and safety of donepezil in the treatment of Alzheimer's disease: An interim analysis of the results of a US multicentre open label extension study. *European Neuropsychopharmacology* 8; 1: 67-75.

Sano M, Ermesto C, Thomas RG, Klauber MR, Schafer K, Grundman M, Woodbury P, Growdon J, Cotman CW, Pfeiffer E, Schneider LS & Thai LJ (1997) A controlled trial of selegiline, alpha-tocopherol, or both as treatment for Alzheimer's disease. *New England Journal of Medicine* 336 (17): 1216-1222.

Skurla E, Roger JC & Sunderland T (1998) Direct assessment of activities of daily living in Alzheimer's disease. *Journal of the American Geriatrics Society* 36: 97-103.

Stebbins GT, Gilley DW, Wilson RS, Bernard EJ & Fox JH (1990a) Effects of language disturbances on premorbid estimates of IQ in mild dementia. *The Clinical Neuropsychologist* 4: 64-68.

Stebbins GT, Wilson RS, Gilley DW, Bernard BA & Fox JH (1990b) Use of the National Adult Reading Test to estimate premorbid IQ in dementia. *The Clinical Neuropsychologist* 4: 18-24.

Sutherland T, Molchan SE & Zubenko GS (1995) Biological Markers in Alzheimer's Disease in Bloom FE & Kupfer DJ (eds), *Psychopharmacology: The Fourth Generation of Progress*, New York, Raven Press, 1389-1399.

Tariot PN (1998) Evaluating response to metrifonate. *Journal of Clinical Psychiatry* 59; Suppl 9: 33-37.

Thompson SBN (1997) *Dementia: A Guide for Health Care Professionals*. Arena, Aldershot.

Wechsler D (1981) *Wechsler Adult Intelligence Scale Revised*. New York: The Psychological Corporation.

Wechsler D (1988) *Wechsler Memory Scale Revised*. San Antonio: Harcourt Brace Jovanovich.

Weinstock M (1999) Selectivity of cholinesterase inhibition. Clinical implications for the treatment of Alzheimer's disease. *CNS Drugs* 12; 4: 307-323.

WSHA-West Sussex Health Authority (1999) *Drug Protocol for Aricept in Alzheimer's Disease*. Referral Guidelines. Worthing: WSHA.

World Health Organisation (1992) *World Health Organisation. The ICD-10 Classification of Mental and Behavioural Disorders: Clinical Description and Diagnostic Guidelines*. Geneva, World Health Organisation.

Zigmond AS & Sniath RP (1983) The Hospital Anxiety and Depression scale. *Acta Psychiatrika Scandinavika* 67: 361-370.

Zirm B, Steinwachs KC, Pracher G & Zirm A (1994) Effekte von Nimodipin und Piracetam auf das kognitive Leistungstempo und die klinische Symptomatik bei

Patienten mit vaskularer und degenerativer Demenz-Ergebnisse einer randomisierten klinischen Vergleichsstudie. [The effects of nimodipine and piracetem on information-processing speed and clinical symptoms of dementia: Results from a randomized clinical trial.] *Zeitschrif fur Gerontolpsychologie und Psychiatrie* 7 (3):157-168.

RECEIVED: 4/22/01
REVISED: 6/22/01
ACCEPTED: 8/2/01

Chapter 8

‘HOLLYWOOD WIVES’: EDUCATION AND SUPPORT FOR SPOUSES OF INPATIENTS WITH DEMENTIA

S.B.N. Thompson & F. Watson

The Journal of Dementia Care (2001), 9 (2), 38.

INTRODUCTION

The impact of a diagnosis of dementia is under-estimated, especially when it is a close relative or spouse (Ramsay, *et al.*, 1995; Thompson, 1997).

A carers group for six wives of hospital in-patients diagnosed with dementia was established with a seven-week programme of 13 four-hour sessions for education and support. A satisfaction questionnaire was administered before and after the programme. A tongue-in-cheek naming

of the group after the TV programme *Hollywood Wives,* was warmly supported!

Topics were proposed and chaired by the authors; some were introduced with a short presentation. Rating scores from questionnaires ranged: 0-5 (definite ‘no’ to definite ‘yes’).

RESULTS

- Three wives felt stress/anxiety due to their husband’s circumstances;
- Three felt sessions helped with stress;
- Five felt the occupational therapist was the most approachable member of the community team (then nurse, doctor). After the final session scores evened out across the team because the wives felt able to approach the most appropriate person;
- Four wives said they felt able to give support to others in a similar position. This dropped to one after the occasion, due to increased awareness of the disease process and a clearer understanding of the professional help that can be obtained;

- Three wives felt more information about their husband's illness and the community services available would have made care at home easier, safer and possible for a longer time. After the sessions four felt information provided in the programme would have met all of these needs, if it had been available during the early stages of illness.

Comments from group members included the following:

- 'Very reassuring to meet and keep contact with wives'…'More beneficial much earlier in my husband's illness'…'a great helping understanding one's own and others' difficulties'.
- Lack of structure of husband's time on ward;
- Inconsistency in care given by some staff;
- Little stimulation or reality orientation throughout the week.

Focus on their husband's needs tended to reduce the importance of their own needs. None had taken a holiday in the past year, irrespective of financial constraints. Some had neglected their own interests due to feeling guilty at leaving their husband behind in hospital.

The group discussed two particular anxieties:

(i) genetics – worries about heredity of Alzheimer's disease were reduced by presentations;

(ii) patient's awareness – 'recognition' is a high level of awareness; not all husbands recognize their spouses but sometimes exhibited 'islands of memory' when visited on the ward.

Focus on the agenda items was important, particularly as the group became more cohesive and vocal. The group was effective in decreasing stress, establishing a support network and providing essential education. It continues to meet.

REFERENCES

Ramsay, M., Winget, C. & Higginson, I. (1995). Review: Measures to determine the outcome of community services for people with dementia. *Age & Ageing*, 24, 73-83.

Thompson, S.B.N. (1997). *Dementia: A Guide for Health Care Professionals.* Arena, Aldershot.

Chapter 9

INTERDISCIPLINARY CLINIC FOR ADULTS WITH EARLY ONSET DEMENTIA IN A MENTAL HEALTH NHS TRUST.

S.B.N. Thompson, T. Coates, F. Chaâbane, P. Cherry,

L. Collins, H. Pennicott, F. Watson & N. Rogan

Clinical Gerontologist (2006), 29 (4), 99-104.

ABSTRACT

A clinic to assess, diagnose and initiate treatment for people under the age of 65 years who have dementia or cognitive dysfunction not attributable to head injury was established for a 6-month trial and operated under the title “Adult Cognitive Assessment Team” (ACAT). This service was established and delivered by professionals from the “older Persons Mental

Health Services" directorate who had particular interests and expertise in this area of care. Evaluation of patient outcome and of the impact of this new service provision informed service providers and planners. The success of the clinic has encouraged this NHS Trust to continue the clinic for a further period and to develop and improve education and consultancy.

INTRODUCTION

The National Service Framework for Older People (NHS, 2001) makes specific reference in Standard 7 to the need to action a review of current arrangements within the National Health Service (NHS) for the management of dementia in younger people. Furthermore, it suggests there is a need to implement local protocols across primary and secondary services. Previous studies have indicated the special issues for younger users of services, particularly those who have an early onset dementia (Luscombe, *et al.*, 1998).

Initially, a survey of all general practitioners (GPs) in the Bognor Regis area (South coast town in West Sussex between Portsmouth in the

West and Chichester in the East) was undertaken to attempt to identify (1) the estimated or actual prevalence of dementia in the younger age group per practice, (2) the current treatment/referral options exercised by GPs in these circumstances, and (3) the specialist service requirements to address the needs of both this patient group and the primary care services. Results of this survey led to the establishment of an "Early Onset Dementia Steering Group" that comprised members of the private and voluntary sectors. This group focused on identifying actual and predicted local needs and, where possible, planning services around those needs.

PROJECT DEVELOPMENT

Lengthy consultation with users, carers, primary care services and secondary care professionals resulted in the formation of a core project group of professionals from Older Persons Mental Health services that have particular interest and expertise in the assessment, diagnosis and treatment of dementia. The recognition of unmet needs within the NHS Trust led to the development of a specialist out-patient clinic for younger people with cognitive dysfunction and/or suspected early onset dementia.

Demographic data showing a population of 175 000 (CPCS, 1997) reflected prevalence within the West Sussex region of approximately 230 patients under the age of 65 years with a known dementia. A brief survey of patients on active caseloads within the Chichester and Bognor Regis Community Mental Health Teams for Older People revealed a total of 23 known patients across the two areas. While this is not a definitive indicator of need, it is probably a fairly accurate predictor of prevalence, taking into consideration that a significant number of people who have dementia in this younger age group will not be known to mental health services as a matter of course. Also, this figure does not take into consideration, for example, the number of patients who are actively known to the Working Age Mental Health Services or to neurology services.

Elsewhere, similar services have been offered fro some time, for example, in London, the Alexandra Service, and also in Cambridge and in Greater Manchester (Baldwin, 2003). However, these services follow consultation with neurology and other professions and do not usually accept direct referrals from GPs or primary care staff. A review of services nation-wide for people under the age of 65 years with cognitive decline not attributable to head injury was limited. While such services

exist, some offering a range of interventions including day care and day hospital places, community support working or networks and continuing care facilities, there was no comparable service against which to objectively benchmark in terms of staffing, facilities, funding and resources and population demography. An eclectic model of service was therefore accepted, based on best practice in established services and resources within the Trust, and developed over the project period.

Memory clinics are established in many areas throughout the United Kingdom (Wright & Lindesay, 1995) as a clinically efficient and cost-effective process for assessing and rehabilitating out-patients who have specific impairment, and may complement organic day hospital provision as an effective alternative to present services, particularly for some out-patients with early onset or mild to moderate dementia (Thompson, 1997; Thompson, et al., 2001) and head injury. While this model of practice is not one that exists within the Trust, they are recommended as good clinical practice in the NHS National Service Framework for Older People (NHS, 2001) and a previous trial of running a memory clinic for people aged over 65 years in 2002 acted as a foundation in terms of experience upon which to base the Adult Cognitive Assessment Clinic (ACAT) service.

Development of this service was additionally guided by information and experience gained from a project which commenced in 1998, for one year, within Older Persons Mental Health Services in the previous Trust (Sussex Weald and Downs NHS Trust) which attempted to establish the effects of Donepezil on visual memory in those patients with mild to moderate Alzheimer's Disease (Thompson, *et al.,* 2001). The project was funded by Pfizer Pharmaceuticals and involved the recruitment of a liaison (CATE – Cognitive Assessment, Treatment and Evaluation) nurse to support the prescription of Aricept to selected patients, the evaluation of assessment tools and the psychosocial impact of treatment with Aricept. The Consultant (in Old Age Psychiatry) and the Consultant Clinical Neuropsychologist for the CATE Project also led the ACAT service.

ACAT CLINIC SERVICE AND STRUCTURE

The ACAT clinic was formed with the aim of responding directly to GP referrals, with the service tailored specifically towards assessment, diagnosis and treatment (where appropriate) of cognitive disorder by use

of domiciliary and outpatient services, using inter-disciplinary expertise in the particular clinical, psychological and social needs of this care group. The clinic is distinct from other NHS regions in providing an inter-disciplinary review meeting prior to each clinic that considers both new referrals and the follow-up of patients seen in the preceding clinics. By developing maximum use of clinic time, patients referred to this service were seen at home within two weeks of referral and in the clinic within four weeks.

The team members included a consultant in old age psychiatry, specialist registrar in old age psychiatry, consultant clinical neuropsychologist, senior occupational therapist, senior social worker, 3 community psychiatric nurses (G grades), clinical nurse specialist, and management and clerical support. Furthermore, supplementary services such as physiotherapists, and speech and language therapy services were involved in the core project development and were accessible as required once the clinic was operational, on a case-by-case basis.

The co-ordination of clinic appointments was undertaken by the manager who acted as the sole referral and entry portal into the project service. Once audit and demographic data had been collated, referrals were discussed at each inter-disciplinary meeting and processed from that

point by the care co-ordinator identified, using the Single Assessment Process as the foundation data format.

A home visit by a clinician from the team enabled personal, social, medical and current history to be taken, in addition to introducing the service and explaining the process that would follow. Subsequent appointments to receive formal assessments from the consultant psychiatrist, consultant neuropsychologist and occupational therapist, spread over 3-4 clinic attendances, ensured that the most appropriate screening and assessment occurred. In addition to these reviews, home visiting continued throughout and the patient was subject to comprehensive physical screening.

Close links were developed with neurology services in the adjacent district general hospital, resulting in the seamless and rapid sharing of patient assessments both to and from this department. Access to MRI and CT scanning was more rapid in comparison to the usual outpatient referral process, with all patients being seen within the time frame of the ACAT clinic duration itself.

Work is underway to develop an education and training component, maximizing on the work already undertaken locally on informing GPs and primary health care teams of the role of the ACAT services, and a

national publication addressing the educational and training needs of GPs (Coates, 1996). In addition, an education project is being proposed that will provide relatives of ACAT clinic attendees with advice on ways of identifying their own risk factors for cognitive decline or change, and practical ways in which their risks can be minimized. This will involve joint working with primary care teams and Primary Care Trusts and is in keeping with current Department of Health initiatives in respect of improving the health of the population.

REFERENCES

Baldwin, R. (ed.) (2003). *Young People with Dementia: A Multidisciplinary Approach.* London: Martin Dunitz.

Coates, T.D. (1996). Dementia. *General Practitioner,* 1 Mar, 55-61.

CPCS – Chichester Priority Care Services NHS Trust (1997). *Welcome to Chichester Priority Care Services.* SH1/25.04.97. Chichester, CPCS.

Luscombe, G., Brodaty, H. & Freeth, S. (1998). Younger people with dementia: Diagnostic issues, effects on carers and use of services. *International Journal of Geriatric Psychiatry,* 13 (5), 323-330.

NHS – National Health Service (2001). *National Service Framework for Older People*. London, Department of Health.

Thompson, S.B.N. (1997). *Dementia: A Guide for Health Care Professionals.* Aldershot, Arena.

Thompson, S.B.N., MacDonald, J. & Coates, T.D. (2001). Improving visual memory with Aricept (Donepezil Hydrochloride, E2020) in mild-to-moderate Alzheimer's disease. *Clinical Gerontologist*, 24 (1/2), 55-73.

Wright, N. & Lindesay, J. (1995). A survey of memory clinics in the British Isles. *International Journal of Geriatric Psychiatry*, 10, 379-385.

PART III: Future Direction (Published Papers)

Chapter 10

TESTAMENTARY CAPACITY AND COGNITIVE REHABILITATION: IMPLICATONS FOR HEAD INJURED AND NEUROLOGICALLY IMPAIRED INDIVIDUALS

S.B.N. Thompson

The Journal of Cognitive Rehabilitation (2009), 27 (Fall), 11-13.

ABSTRACT

Many married couples have their property owned as joint tenants. Upon the death of either person, ownership of the estate automatically passes to the survivor. However, if the property is owned as tenants in common, then the one half of the estate belonging to the deceased is dealt with by their will. Problems arise when there is no will, when others make a claim, or when another will is executed.

INTRODUCTION

Testamentary capacity is the legal term used, in common law tradition, to describe a person's legal and mental ability to make a valid will. There are three premises: (i) Presumption of capacity, (ii) Requirements, and (iii) Proof of testamentary capacity.

(i) *Presumption of capacity*

There is often dispute over a person's possessions and estate if the individual has died without leaving a valid will or if the will is question. Problems may also arise when a second or subsequent will has been found that contradicts the first or previous wills or is unusual in some way. It is particularly difficult when the owner of the estate has had enduring neurological problems together with mental difficulties. People with head injuries and progressive neurological diseases can be victims of unscrupulous persons prior to passing on their estate simply because of their vulnerability and unprotected status.

Litigation in this respect typically revolves around the lack of mental capacity to make a will. The will presented or executed is challenged by

others if they know that the person making the will did not know what s/he was doing when they executed the will.

This law applies to adults since minors are conclusively deemed incapable of making a will by the common law. Exceptions are minors who serve in the military who are conceded the right to make a will by statute in many jurisdictions.

I was asked recently to help investigate a case in which the will-maker suffered from early onset Alzheimer's disease and had also suffered a head injury. Since a second will was also executed that substantially contradicted the first will, the potential inheritors contested the second will suggesting that someone had persuaded the vulnerable person to change his will and that he did not have testamentary capacity at the time of the second will's execution. This case poses several difficult questions for others facing this situation. Namely: (1) Are all parties involved unbiased in the decision over whether or not the will-maker was able to make a valid decisions about his estate? (2) Are there sufficient documents and evidence such as hospital notes and physician's notes to satisfy the enquiry? (3) Is there any other evidence to support or refute the allegations such as a letter or event that demonstrates mental ability or a lack of ability?

(ii) *Requirements*

While the wording of statutes or judicial rulings often vary from one jurisdiction to another, the requirements for testamentary capacity are fairly minimal. The test generally requires that the testator (or will-maker) was aware of the following:

- the persons who are the natural beneficiaries;
- the extent and also the value of their property and estate;
- the disposition that they are making by executing the will;
- how this action affects the distribution of the estate.

The burden of proof is on the party propounding the Will to show that the testator did have the relevant capacity (Dukeminier & Johanson, 2005).

(iii) *Proof of testamentary capacity*

The challengers of the will must show that that the will-maker suffered from some sort of mental unsoundness that rendered them unable to remember family members or caused them to hold insane delusions about

them. Difficulty arises when the will-maker may have had periods of lucidity and but also had occasions when they were unsure of their surrounds or who was with them. It is typical in some progressive and deteriorating neurological diseases and in head injury for a person to suffer, on occasions, loss of memory or “islands of memory” when they can and cannot recall events or people (Thompson, 2006). Establishing what actually happened at the time of a revised will may take a considerable time and can be dependent upon helpful parties as well as sufficient and useful documentation. The process can also be hampered when concerned parties consider there to be a lot at stake in terms of financial gain or profit.

Indeed, on many occasions the Court have found that whilst a testator has been found generally to lack testamentary capacity due to infirmity, senility or insanity, sometimes they will still find that the testator has had a temporary period of lucidity. Therefore, the will stands as valid.

DISCUSSION

Isaac Ray (1807-1881), often regarded as the father of American forensic psychiatry, proposed that there was a role for a forensic psychiatrist in a will contest. The psychiatrist has relevant knowledge to analyze available information in determining competency (Ray, 1838).

While some patients with Alzheimer's disease can be relatively stable and unimpaired for years, some patients gradually become more cognitively impaired over time (Tabert, Manly, Liu, Pelton, Rosenblum, Jacobs, Zamora, Goodkind, Bell, Stern & Devanand, 2006). Hence, there may be a need to intervene in circumstances where the executor of the will may have been vulnerable to other influences and where the will has been changed.

An interesting case is the one cited by Bursztajn (1992). "R" and "M" began dating in 1968. Although they never married, R and M lived together in a house owned by R for the last 14 years of their relationship. In 1996, R executed a deed to his house, leaving it to M upon his death. In 1999, R began to suffer from Alzheimer's disease.

In May 2002, R's daughter took R to a lawyer's office where he changed the deed to his house to leave it to his daughters rather than to M.

No forensic evaluation was conducted to assess his testamentary capacity. M sued, claiming that R was not competent in 2002. However, the daughters claimed that R made a conscious decision that he wanted the family home to stay in the family. At a pre-trial motion, the court ruled that while a deed is usually a contract, under these circumstances it would be considered as a gift. The premise is that much less mental ability is required to make a valid gift than to make a contract.

This was not the end to the case. Dr Busztajn was asked to conduct a forensic psychiatric evaluation. Medical records revealed R's progressive cognitive loss during the years between 1999 and 2002 and pre-trial discovery revealed that by 2002, R did not know the day or the year, the fact that he was retired, and was unable to prepare food or use the telephone. He had also lost his ability to think abstractly.

Sadly R died before trial. At trial, R's daughter insisted that her father exercised his own free will when he changed the deed to his house. However, under cross-examination, further evidence emerged concerning her father's lack of mental ability, concluding in the Defendants agreeing to make a financial settlement with M.

CONCLUSION

The law is designed to protect individuals so that they can exercise their right to pass on estates after death. It is also intended to protect beneficiaries. Difficulty arises when it is no longer clear what the individual's intentions have been and whether or not their actions have been executed with free will and with full intellectual capacity.

The law continues to struggle with the question of how to measure testamentary capacity effectively. Champine (2005; 2006) has proposed that the law ought to allow testators alternative means of satisfying the testamentary capacity standard. There should be an option to validate a testator's capacity during their lifetime through use of a forensic assessment instrument that measures the cognitive elements of testamentary capacity.

Whilst this would certainly help those individuals with a progressive disease, it still does not address satisfactorily those who have suffered a specific and discrete event such as a head injury. It does not remove the difficulty of knowing the status of person at a specific time line; rather it goes some way to describing a person during their lifetime in terms of

mental ability and capacity. It is perhaps time to re-think the standard of testamentary capacity and how it can be met. After all, contract law has been struggling with this for more than a century.

REFERENCES

Bursztajn, H.J. (1992). Competency to make a will. *American Journal of Psychiatry* 149: 1415.

Champine, P.R. (2005). A blueprint for testamentary capacity reform. *Social Science Research Network* ID696081_code300961.pdf, Mar 31: 1-2.

Champine, P.R. (2006). Expertise and Instinct in the Assessment of Testamentary Capacity. *Villanova Law Review* 51: 25-93.

Dukeminier, J. & Johanson, S.M. (2005). *Wills, Trusts & Estates.* 6th ed, Aspen, New York.

Ray, I. (1838). *A Treatise on the Medical Jurisprudence of Insanity*. Little, Brown & Co., Boston.

Tabert, M.H., Manly, J.J., Liu, X., Pelton, G.H., Rosenblum, S., Jacobs, M., Zamora, D., Goodkind, M., Bell, K., Stern Y. & Devenand, D.P.

(2006). Neuropsychological prediction of conversion to Alzheimer disease in patients with mild cognitive impairment. *Archives of General Psychiatry* Aug; 63; 8: 916-924.

Thompson, S.B.N. (2006). *Dementia and Memory: A Handbook for Students and Professionals.* Ashgate, Aldershot.

PART IV: In Summary

Chapter 11

DISCUSSION

This chapter is subdivided into:

- Support and refutation of hypotheses.
- Suggestions on possible methodological improvements.
- Summary and conclusions of empirical studies.
- Future direction of research and key to improving services.

11.1 SUPPORT AND REFUTATION OF HYPOTHESES

Social abilities

It was hypothesised that dementing individuals with Down's syndrome show greater decline in social abilities with age, compared with other groups of dementing people with learning disabilities.

On comparing data obtained from the 12-month studies from both client groups, a statistically significant difference was found in the 'sum of social "difference" scores' of the DMR after 12 months ($p < .002$). Although the majority of HADS scores for clients fell within the 'normal' range (ie below 8 points), the effect of controlling for these scores was evidenced when they were partialled out in data analyses: $p < .02$ (after HADS Anxiety was partialled out), and $p < .003$ (after HADS Depression was partialled out). Therefore, the level of statistical significance of this finding was reduced following controlling for the HADS scores. The Down's syndrome clients showed a greater score difference between assessments 1 and 2 in the 'sum of social scores' than the non-Down's syndrome clients. This supports hypothesis 1. There was a greater variance in the scores of the Down's syndrome clients.

It is known that people with Down's syndrome have developmental delays (Maclean, *et al.*, 1991) and very often also have expressive and receptive (Young & Kramer, 1991) language difficulties. Therefore, it is proposed that the differences in scores between people with Down's syndrome and other learning disabilities may be at least due partly to the known differences in understanding language and in the ability for expressive language. Since language is important in the development of

social abilities, this impediment may well account for the difference in social abilities seen between the two client groups.

The finding that the Down's syndrome clients showed a greater score difference between assessments 1 and 2 on this measure is interesting in light of reports that suggest Down's syndrome people have neuropsychological profiles similar to those who have been diagnosed with Alzheimer's disease (Deb & Braganza, 1999). Such clients have also shown signs of declining social skills. Other reports support the view that changes in a person's personality, such as sociability and social skill performance, is affected in people with Down's syndrome who have been diagnosed with Alzheimer's disease (Evenhuis, 1992a; 1997).

In comparison with Evenhuis' (1992a,b; 1996) samples, for both client groups in this study the means of DMR 'sum of social scores' were higher; therefore, they had a better range of social skills to begin with (as indicated by their DMR scores) and were generally more able. Yet, it is interesting to note that 15 clients from the two groups of the 12-month studies showed a significant decline in these scores between assessments 1 and 2 (Thompson, 1999; 2000a).

Eighteen clients in the Down's syndrome group and 19 clients in the non-Down's syndrome group of the 12-month studies showed changes in

their DMR 'sum of social scores' between assessments 1 and 2. Of the Down's syndrome clients, nine fulfilled Evenhuis' (1992b) criteria for the diagnosis of a dementing process, ie they had a 4-point score change between assessments. Of the non-Down's syndrome clients, six showed a 4-point score change. Evenhuis (1992a) includes a further set of criteria related to IQ level. For the "high-moderate level of retardation (IQ = 45 - 55)", the change in 'sum of social scores' is given as greater than or equal to 15. Three clients from the Down's syndrome clients (DS8, DS13 and DS14) fell into this category. The mean IQ of the non-Down's syndrome clients was slightly higher than for the Down's syndrome group; therefore, Evenhuis' (1992a) "mild-moderate level of retardation (IQ = 55 - 70)" applies where the change in 'sum of social scores' is given as greater than or equal to 10. In the non-Down's syndrome group, one client (NDS16) fell into this category. Overall, the Down's syndrome group had <u>more</u> clients who had <u>greater</u> changes in their 'sum of social scores' between assessments 1 and 2 (Thompson, 1999).

In the 6-month study (Thompson, 2002), no evidence was found to suggest that the Down’s syndrome clients had greater social score changes than the non-Down’s syndrome group. It may be that any differences between the two groups does not become significant until

after a longer period of time, ie 12-month interval between assessments.

It was further hypothesised that social changes co-vary in dementing individuals with Down's syndrome, as compared with non-Down's syndrome individuals. In the 6-month study (Thompson, 2002), analysis of within-group data revealed social score changes of only the Down's syndrome group were significant ($p < .005$). Hence, supporting this further hypothesis.

Examination of 12-month data for each client group individually, revealed that for the Down's syndrome group, there was a significant difference between assessments 1 and 2 on the DMR 'sum of social scores' ($p < .001$). A significant difference remained following the partialling out of HADS scores, again supporting this further hypothesis. These scores were not significant for the non-Down's syndrome group. Hence, this is strong evidence for suggesting that social abilities significantly co-vary over time in people with Down's syndrome but not for those in the non-Down's syndrome group.

Examining linear associations between measures

It does seem to be the case that cognitive abilities (as measured by the

DMR 'sum of cognitive "difference" scores') are not associated in a linear manner with scores on the MEAMS as no significant linear relationship was found for data of either client group. However, a significant (though weak) linear relationship was found between the DMR 'sum of social "difference" scores' with the MEAMS number of subtests passed “difference” scores ($r = .522$; $p < .04$; 1-tailed; after controlling for HADS scores) for the Down's syndrome group. Hence, it is suggested that if social abilities decline in people with learning disabilities (particularly those with Down's syndrome) then this decline may be correlated with their performance on the MEAMS.

As this is distinct from clients in the non-Down's syndrome group, this lends further support for the hypothesis that suggests social abilities decline over time in people with Down's syndrome as compared with people with non-Down's syndrome learning disabilities.

Cognitive abilities

It was hypothesised that dementing individuals with Down’s syndrome show greater decline in cognitive abilities with age, compared with other groups of dementing people with learning disabilities.

However, no significant difference was found when comparing the two client groups on the DMR 'sum of cognitive scores' over time points. Hence, this hypothesis was refuted.

A further hypothesis proposed that cognitive changes co-vary in dementing individuals with Down’s syndrome, as compared with non-Down’s syndrome individuals. In the 12-month studies, a significant score change was found ($p < .001$) in the Down's syndrome clients suggesting that cognitive abilities co-vary over time, supporting this further hypothesis. Data from the non-Down's syndrome clients also supported the view that cognitive abilities (as measured by the DMR 'sum of cognitive scores') decline over time ($p < .05$; HADS Anxiety partialled out). Although, when HADS Depression was partialled out, these scores were not significant.

It is possible that some of these clients suffered from mild depression (whether or not as a consequence of dementia) and hence their performance on tests of cognitive abilities were affected.

In the 6-month study (Thompson, 2002), significant score changes in cognitive abilities were found for both the Down’s syndrome group (MEAMS Number of Subtests passed: $p <.005$) and also for the non-Down’s syndrome group (MEAMS Total score: $p <.01$; MEAMS Number

of Subtests passed: $p < .005$). Hence, this hypothesis was refuted for the 6-month time interval.

It may well be that decline in social abilities occurs before decline in cognitive abilities, or possibly that the latter is more subtle and difficult to measure in these clients. A decline in social skills, for example, may be more noticeable and easily assessed because of the person's withdrawal from socialising or because of decreased interaction with care staff (Thompson, 1993; 1997). Little is known in the learning disability population about the rate of decline (Thompson, 2002), or whether decline in cognitive abilities occurs in the same way as it seems to occur in people without learning disabilities who are dementing; although, rate of cognitive decline with age in people who are not dementing who have Down's syndrome or "other" learning disabilities seems to be indistinguishable from one another (Das, *et al.*, 1993; Pulsifer, 1996; Vincent, 1996).

Comparing findings from the 6-month study with the 12-month studies, it would seem that the rate of decline in abilities may be different in the two client groups. A greater decline in cognitive scores was evidenced in the Down’s syndrome group over a longer 12-month period.

Golding and others have discussed the use of the MEAMS dementia

screening tool (Golding, 1989; van Belle, *et al.*, 1990). In the MEAMS manual, Golding states that it systematically surveys the major areas of cognitive performance, namely, orientation, name learning, comprehension, remembering pictures, arithmetic, spatial construction, fragmented letter perception, unusual views, usual views, verbal fluency, and motor perseveration. Although she states in her three-level criteria, score performance, she does not report test-retest data using the parallel version of the test. Hence, the actual amount of discrepancy expected between tests is not known. Indeed, the degree of discrepancy seems to be very dependent on the population studied (Powell, Brooker & Papadopolous, 1993; Dickson, *et al.*, 1995).

For the Down's syndrome group, only two clients (DS11 and DS12) scored above 7 (number of subtests passed) at assessment 1. At retest (assessment 2), no clients scored above 7; hence, according to Golding (1989) this would place all these clients in the third (lowest) category where 'more detailed investigation' is recommended. The other two categories are stated as: score 10 - 12 (normal); and scores 8 or 9 (borderline range). It is curious to note the slight improvement in some scores though all scores were still below the 7-point cut-off stated by Golding (1989) with a mean score for the "differences" between

assessments 1 and 2 (for all Down's syndrome group data) of only 0.30, and a very large variance (standard deviation = 13.33).

For the non-Down's syndrome group, the mean of scores for the MEAMS number of subtests passed was higher than for the Down's syndrome group with a mean for these "difference" scores of -0.57 and a lower variance and less spread than for the Down's syndrome group (standard deviation = 3.51). Although no statistical difference was found between the two client groups on the MEAMS number of subtests passed, it would seem that of the two client groups, more clients in the non-Down's syndrome group showed a deterioration in their performance on the MEAMS than did the Down's syndrome group. It is possible that cognitive abilities were more developed in the non-Down's syndrome clients and may have been more prone to the debilitating effects of dementia.

Cognitive impairment is salient in dementia in the non-learning disabled population (Dickson, 2001). In particular, executive phenomena have received recent interest (eg Baddeley, 1998; Greene, Hodges & Baddeley, 1995), which have led to the development of a new assessment measure, the Behavioural Assessment of the Dysexecutive Syndrome (Wilson, *et al.*, 1996).

Shallice (1988) and Furster (1993) have also explained the cognitive impairment in dementia in terms of a failure in sequencing and executive phenomena, such as the ability to organise and plan, and Morris (1996) has described this in terms of a 'dysexecutive syndrome'. Yet, there is still a lack of substantive evidence of such similar impairments in people with learning disabilities who have dementia, although it is thought that people with Down's syndrome tend to decline cognitively (Das, *et al.*, 1993), but the rate of decline seems to vary considerably. Neither has this been particularly well documented, nor have there been well-matched comparisons with non-Down's syndrome learning disabled people with a diagnosis of dementia. Clearly, there is scope for further research in this area, in particular, investigating whether or not there is a breakdown in the Articulatory Loop System (ALS), one of the supporting systems of Baddeley’s (1986; 1998) Central Executive System (CES).

Indeed, the cognitive impairment seen in people with dementia may also be considered in terms of Baddeley's (1986; 1998) Central Executive System. A breakdown in the ALS, responsible for recycling verbally encoded information, may account for the difficulties that learning disabled people (particularly Down's syndrome) tend to have with tasks that require intact verbal memory functioning. Language difficulties also

impact on tests of cognitive functioning that require verbal responses, for example, the 'verbal' subtests of the WAIS-R and the word fluency subtest of the MEAMS. However, despite some reports of articulatory rehearsal impairments in Alzheimer's disease (eg Hulme, Lee & Brown, 1993), impressive evidence comes from 'articulatory suppression' tasks which tend to suggest that perhaps the ALS is not implicated in dementia since, particularly Alzheimer-type patients, perform equally well to normal controls. This finding may lend support to the lack of a significant difference in this study between the two client groups in their MEAMS scores.

In the visuospatial domain, the visuospatial sketchpad (Baddeley, 1986; 1998), which maintains visuospatial imagery, is thought to be operative in tasks requiring spatial construction (Sahakian, *et al.*, 1988); to some extent, this applies to the visuospatial subtest of the MEAMS. Visuospatial ability is thought to be significantly impaired in people with dementia; however, it has not been investigated extensively (Morris, 1994a; 1996) and therefore, is another are for future invetigation, especially in the learning disability population.

Problems of a differential diagnosis

In terms of the HADS scores between assessments 1 and 2, it was found that both HADS Anxiety and HADS Depression scores co-varied over time, though more significantly in the Down's syndrome group. This finding can possibly be explained in terms of people's frustrations and low mood which has been evidenced in the non-learning disability population who are dementing (Mohanarubin, Sastry & Finucane, 1989; Lamberty & Bieliauskas, 1993).

However, it is not known whether the presence of depression is more salient in people with Down's syndrome versus other learning disabilities with or without dementia. Although, it is known that depression is one of the factors that may impact on a differential diagnosis of dementia in both the normal population (De Groot, *et al.*, 2000; Dickson, 2001; Krishnan, 1991) and in Down’s syndrome (Evenhuis, 1997). It is important here to address the problem of a differential diagnosis (see Rossor, 1999), particularly between dementia and depression (O’Brien, *et al.*, 1996). On examining HADS scores for both groups across all assessments, few clients' scores were found to be in the borderline range with the majority falling into the 'normal' category. It is recognised that discriminating

between dementia and depression in the non-learning disability population is difficult but not impossible (Krishnan, 1991; Krishnan & McDonald, 1995; Krishnan, Hays & Blazer, 1997; Rao, 1998), for example, using the Beck Depression Inventory (Beck & Steer, 1987) in conjunction with measures of social and cognitive abilities and the clinical interview.

However, in the learning disability population, it is increasingly difficult when language and comprehension is limited. Despite these difficulties, it is believed that the modified use of the HADS (in an interview situation) and the fact that third parties (namely, carers) also contributed to the data pool (by completing the DMR, for example), helped to strengthen the methodology implemented, and hence produce valid results. In addition, biographical data helped to confirm that none of the clients had been observed to be depressed prior to the study nor did any of them have a known history of depression or low mood swings.

11.2 SUGGESTIONS FOR POSSIBLE METHODOLOGICAL IMPROVEMENTS

Criticising any study in terms of the methodology used is important not only in order to interpret findings hopefully more accurately but also to learn from mistakes and to direct future research. Choosing suitable assessment tools that can be used with people who have learning disabilities is difficult because of limitations of the tools themselves and because of the generally low level of intellectual abilities in this population.

Using the Wechsler Adult Intelligence Scale - Revised (WAIS-R) for example, has to be considered in terms of 'floor effects' (see Crayton & Oliver, 1993). As discussed earlier, sometimes the range of sensitivity of a test does not permit for a 'true' measurement to be taken for a given subject. In the case of a floor effect, the lower limit (or score obtained) may in fact reflect a range of possible values if the scale had extended further. For example, a client may score 2 on a scale ranging from 1 to 5, but if a new scale of 3 to 12 is used where level 3 is re-assigned 1 and 12 re-assigned 10, then the same client's performance can only be recorded as 1 since the scale does not reach beyond this lower point. Hence, this

client would be given an distorted high score. Similarly, use of the WAIS-R has been criticised (Crawford, Gray & Allan, 1995), particularly when used with people with learning disabilities (Atkinson, 1991a). Fortunately, the range of fullscale IQs of the clients in this study fell in the range 45 - 63 (mean of 51.90; standard deviation of 1.21) for the Down's syndrome group, and 51 - 69 (mean of 59.19; standard deviation of 1.17) for the non-Down's syndrome group. None of these clients' scores reached the "floor" of any of the WAIS-R subtests.

However, a similar problem can also apply when a range of scores are clustered around the lower end of a scale - hence, the scale used is not discriminating between clients that would otherwise score lower or in-between the scores on the scale used (ie the scale is not "fine" enough). Clients scores on the WAIS-R were certainly towards the lower end of the subtest scales; however, consistently low performance scores were also achieved on another test of intellectual capacity, the Raven Coloured Progressive Matrices (RCPM). This latter assessment tool is a measure recognised for its suitability and sensitivity for people with low intellectual ability (Raven, Court & Raven, 1990; Vodegel-Matzen, van der Molen & Dudink, 1994), thus tends to confirm the generally low intellectual capabilities of the clients studied.

Testing specific cognitive abilities is also made difficult in people with limited abilities. The MEAMS has been found to be useful in discriminating between depressed and dementing older patients (Golding, 1989; van Belle, *et al.*, 1990), but this has not been verified in the learning disability population. Possible floor effects may well account for the apparent lack of change in some clients' scores between assessments; likewise, apparent improvement or deterioration in scores might be masking a "true" change in performance scores.

In the case of apparently small changes in clients' scores, it is also possible that the MEAMS may lack sensitivity in what it actually measures, or may simply not have sufficient range of scores spanning the item to be measured. Indeed, some changes in cognition may be subtle, but nonetheless they remain a salient indicator of a deteriorating condition in the normal population (Dickson, 2001). In terms of items purported to be measured by the MEAMS, these subtests cover the range of abilities that seem to decline in older dementing adults. For example, the orientation subtest, delayed identification task (similar in some respects to the 'delayed matching to sample task' of Sahakian , *et al.*, 1988), and visual construction item of the MEAMS tap into abilities reported to decline in dementing adults (Baddeley, *et al.*, 1986; Braekus, 1995; Myers

& Goate, 2001). Hence, it was regarded as being an important assessment tool to include in the study, despite some of its forthcomings.

On a statistical note, there is always the possibility of making a type I error (ie rejecting the null hypothesis when it is in fact true) or a type II error (ie accepting the null hypothesis when it is not in fact true) – see Greene and d'Oliveira (1982). The main implications for this study would be:

(i) to state that there is a difference in cognitive and social abilities between people with dementia and Down's syndrome versus non-Down's syndrome, when in fact there is no difference (type I error). This seems unlikely given that evidence is available suggesting people with Down's syndrome may have a predisposition to Alzheimer's disease over non-Down's syndrome groups (Cooper, *et al.*, 2001; Schupf, *et al.*, 2001).

(ii) state that cognitive and social abilities of people with learning disabilities co-vary with time when in fact they do not alter (type I error). There is nothing in the research literature to suggest that this population is any different to the general population in terms of symptoms of dementia. Furthermore, people from all populations diagnosed with dementia seem

to share the same neuropathology for dementia (Grober, Dickson & Sliwinski, 1999; Kawas & Katzman, 1999; Dickson, 2001) and some features of the neuropsychological profile (Spinnler & Dela Sala, 1988) though there is predominantly a lack of substantive evidence about the profile of declining cognitive functioning in people with learning disabilities (particularly non-Down's syndrome).

(iii) state that there are no (or only weak) linear associations between the data when in fact there are (strong) associations (type II error). This is a possibility; however, only testing with even larger samples over a longer time period would confirm or refute these particular assertions.

Making type I and type II errors are calculated risks. The findings of this study tend to support the findings of other studies (eg Evenhuis, 1992b) that show the level of functioning in people with learning disabilities co-varies with time. This study has also revealed differences between Down’s syndrome and other types of learning disabilities with respect to changes in cognitive and social abilities as well as the rate of decline of dementia.

What this study cannot show is the distinction between vascular

dementia and other dementias. Computerised tomography scans also fail to show this distinction (Shapiro, Haxby & Grady, 1992; Markesberry, 1998; Dickson, 2001). Although some authors have extensively described different dementias (eg Lishman, 1987; Miller & Morris, 1993; Litvan, Grimes & Lang, 2000; Dickson 2001), there have been few longitudinal matched studies comparing the different categories of dementia and their course over time. It is acknowledged that further assessments even beyond the twelve-month interval may well have yielded additional and interesting results.

Future research

It was not until 18 May 2000, following the efforts of many researchers, that the historical step was reached, when the complete DNA sequence of human chromosome 21 was determined and published in Nature (Hattori, *et al.*, 2000). The general screening of populations by examining past medical and psychological histories is not a new concept (eg Cooper and Bickel, 1984) with a few promoting the use of routinely screening populations for risk factors for Alzheimer's disease (Milne, 2010).

However, some researchers err on the side of caution and believe that

more knowledge is needed with respect to the progression of the disease since it is very much variant amongst sufferers (Ruitenberg, Kalmijn & de Ridder, 2001; Ruitenberg, Ott & van Swieten, 2001). It is perhaps wise to be cautious especially since researchers have found new links with the disease which put forward another set of risk factors to consider. For example, Velayudhan and colleagues (2010) have found a significant risk of developing dementia amongst people with diabetes. The researchers found that diabetes mellitus is associated with cognitive dysfunction and also with the progression of Alzheimer's disease.

There has been considerable advancement in our knowledge of Alzheimer's disease, particularly in genetics, (eg Bertram, Blacker & Crystal, 2000; Myers, Holmans & Marshall, 2000; Pericak-Vance, Grubber & Bailey, 2000; Scott, Grubber & Conneally, 2000). However, there is still some way to go before we fully understand the link between Down's syndrome and Alzheimer's disease and why some people with Down's syndrome do not have Alzheimer's disease despite their genetic predisposition (Pinter, *et al.*, 2001; Schupf, *et al.*, 2001; Cooper, *et al.*, 2001).

It is hoped that further discoveries will help improve management and treatment approaches for sufferers in both the normal population and for

people with Down's syndrome. In the United Kingdom, the National Institute for Clinical Excellence has published guidelines for the treatment of Alzheimer's disease in people with learning disabilities (Arshad, Sridharan & Brown, 2001) which is a giant step compared with decades of unspecific approaches that have tended towards management rather than treatment, *per se* (Thompson, 1993; 1997; 2000b). The link between vascular dementia, Alzheimer's disease and stroke has also meant that treatment may be advanced (Rao, 1998; Rhodin & Thomas, 2001).

Such advancement of theories has included a re-kindling of the neuroplasticity theory (Arendt, 2001) which suggests that mechanisms of molecular and cellular control of neuronal differentiation and proliferation might be critically involved in the pathogenesis of neurodegeneration and recovery. This theory has been advanced as an explanation of peole recovering from strokes (Thompson & Berry, 1997; 1998) and in respect to people with learning disorders (Bigler, 1992).

In 2010, Deng, Aimone and Gage proposed that the integration of adult-born neurons into the circuitry of the adult hippocampus suggests an important role for adult hippocampal neurogenesis in learning and memory. In particular, computational studies suggest that, before newborn neurons are fully mature, they might function as a pattern

integrator by encoding different events that occur closely in time. This might explain the possibility of some neurons re-routing during plasticity after insult or injury.

Media coverage of the famous, for example, ex-President of the United States, Ronald Reagan (Barrett, 1997), often re-kindles public interest in areas of medical and psychological study. It is unfortunate that the area of learning disabilities (and Alzheimer's disease) is generally followed by the public with less interest unless it is discussed together with the 'normal' population, or in light of a celebrity tragedy. Perhaps this situation may change as our older population increases.

11.3 SUMMARY AND CONCLUSIONS OF EMPIRICAL STUDIES

Clients with Down's syndrome (DS) and clients with non-Down's syndrome (NDS) learning disabilities were assessed using specially selected neuropsychological assessment tools at two time points separated by either six or twelve months. Evidence was found to support hypothesis 1 which suggested that people with DS show a greater decline in social

abilities with age, compared with other groups of people with learning disabilities. Statistically, score changes reflecting the social abilities of the DS clients were found to be significantly greater ($p < .002$) than those of the NDS clients.

Evidence was found to support those hypotheses that suggested social and cognitive changes co-vary over time in these two older populations. Scores reflecting the social abilities (DS) and cognitive abilities (DS & NDS) co-varied over time ($p < .001$) with a significant decline in social abilities ($p < .001$) for the DS clients. In the 6-month study, only the Down's syndrome group declined in social abilities ($p <.005$); both groups declined in cognitive abilities (Down's syndrome: $p <.005$; Non-Down's syndrome: $p <.01$; $p <.005$).

A weak linear association ($p < .004$; $r = .625$; 2-tailed) between cognitive performance and social abilities was found for the DS clients. Findings were explained in terms of poor language abilities in DS people generally, the link between declining social abilities and dementia, and Baddeley's (1986) Central Executive System and the dysexecutive syndrome discussed by Shallice (1988) and Furster (1993).

A statistically significant difference ($p < .002$) was found in the 'sum of social scores' on the Dementia Questionnaire for Mentally Retarded

Persons - DMR (ie "social abilities") between Down's syndrome and non-Down's syndrome clients between assessments 1 and 2. This decline was greater in the Down's syndrome clients. No significant difference between client groups was seen in their 'sum of cognitive scores' (DMR) - ie "cognitive abilities". Hence, partial support was found for hypothesis 1, which proposed that the Down's syndrome clients showed a greater decline in (cognitive and) social abilities with age, compared with other groups of people with learning disabilities.

Several explanations were proposed for the differences found between the two client groups; namely, the well-documented difficulties with language (comprehension and expression) in people with Down's syndrome may affect their social abilities. Also, in comparison with Evenhuis' (1992a,b) samples, decline in social abilities has been found to be associated with a dementing process particularly in people with Down's syndrome.

No statistically significant evidence was found of a difference in cognitive abilities over time between client groups though from visual analysis it appeared that more of the non-Down's syndrome clients deteriorated in their performance on the Middlesex Elderly Assessment of Mental State (MEAMS). Hence, the hypothesis was refuted which

suggested that Down's syndrome clients showed a greater decline in cognitive abilities with age, compared with other groups of learning disabilities.

Social and cognitive abilities tended to co-vary over time in the Down's syndrome clients whilst cognitive abilities tended to co-vary in the non-Down's syndrome clients. These findings were discussed in terms of the 'dysexecutive syndrome' (Shallice, 1988; Furster, 1993; Wilson, *et al.*, 1996) which suggests that cognitive impairment in dementia occurs because of a failure in sequencing and executive phenomena, such as planning and organising. Impairments possibly due to changes in visuospatial ability were also discussed in terms of Baddeley's (1986; 1998) 'visuospatial sketchpad' which maintains visuospatial imagery as part of the Central Executive System.

Data were also examined for linear associations. Although a significant association was found between the MEAMS number of subtests passed “difference” scores and the DMR 'sum of social "difference" scores' ($p < .05$), this correlation was weak ($r = .5219$). It was suggested that declining social abilities of Down's syndrome clients may be weakly associated with performance on the MEAMS.

Finally, it is suggested that further longitudinal studies should be

conducted that compare different forms of dementia, and particularly with vascular disorders such as vascular dementia and stroke.

11.4 FUTURE DIRECTION OF RESEARCH AND KEY TO IMPROVING SERVICES

For some time, the establishment of memory clinics within public sector settings such as the National Health Service have been seen to be the way forward for the provision of dementia services (Wright & Lindesay, 1995). However, there is an inconsistency in the approach adopted across the United Kingdom. This has been for a number of reasons. For example, some clinics have ceased after funding has run out; there have been under-representations of key personnel in the interdisciplinary team (eg clinical psychologists or occupational therapists) due to lack of recruitment or retention; and there has sometimes been a lack of clarity in the focus of these clinics.

This latter point is worthy of expansion. Sometimes the origination for the clinic has been to address the over-subscription and demands put upon dementia services. Attendees are not necessarily best served by a

"one-stop" approach if their treatment or support needs are complex and are not adequately followed up. However, interdisciplinary clinics have the benefit for the patient attendee of being able to see all professionals responsible for dementia care in one day. There is a greater understanding of attendees' needs when professionals are able to discuss their patients at the end of the clinic and this often leads to better follow-ups in terms of appropriate care. This only works though when the clinic is properly funded.

In terms of psychological support, clinics involving the carer as well as the patient have greater success in terms of attendance and the psychological welfare of the patient. Increasingly, service providers are canvassing and involving carers in decision making and the development of services for people with dementia. This is so important when the carer often knows the person with dementia better than the service professional. Researchers for some time have become to recognise and accept this fact and are involving carers in such activities as the development of assessments as well as studying the burden put upon carers of people with Alzheimer's disease (eg Given, *et al.*, 2007). It is also refreshing to see that public policy is beginning to acknowledge deficits in mainstream services and to take account of the special needs of people with learning

disability and dementia (eg Janicki, *et al.*, 2007).

However, legal provision for people with dementia is a neglected area and the subject of Will provision is usually only tested with a solicitor or after the person has died. Therefore, this important area needs far more consideration especially as there is a growing population of older people worldwide and an increasing population of people with dementia.

Life-long assessments are being considered as a way forward for people with a diagnosis of dementia (Thompson, 2009). In this way, the deterioration of the patient in terms of cognitive and social abilities can be monitored and addressed from a service provision point of view as well as from the legal perspective. If the cognitive profile of a dementia patient can be captured then it is likely to be useful if their testamentary capacity is contested in court when a legal Will is considered. Whilst no one wishes to consider such aspects during a person's lifetime, there is a legal responsibility of the welfare state to provide advice on such matters so that the growing number of people facing this situation can receive some sort of psychological comfort and protection.

REFERENCES

Agbayewa, M.D., Weir, J., Tuokko, H. & Beattie, L. (1991), 'Depression in dementia: Its impact on functional ability', *Dementia, 2:* 212-217.

Arshad, P., Sridharan, B. & Brown, R. (2001), 'Treatment for Alzheimer's disease in people with learning disabilities: NICE guidance', *British Journal of Psychiatry, 179:* 74.

Atkinson, L. (1991a), 'Mental retardation and WAIS-R difference scores', Journal of *Mental Deficiency Research, 35:* 537-542.

Baddeley, A.D. (1986), *'Working Memory'*, Oxford: Oxford University Press.

Baddeley, A.D. (1998), 'The central executive: A concept and some misconceptions', *Journal of the International Neuropsychological Society, 4:* 523-526.

Baddeley, A.D., Logie, R., Bressi, S. & Della Sala, S. (1986), 'Dementia and working memory', *Quarterly Journal of Experimental Psychology, 38A:* 603-618.

Barrett, L.I. (1997), 'Reagan's long goodbye', *Time, March 24:* 48.

Beck, A.T. & Steer, R.A. (1987), *'Beck Depression Inventory. Manual'*,

San Antonio: Harcourt Brace Jovanovich.

Bertram, L., Blacker, D. & Crystal, A. (2000), ‘Candidate genes showing no evidence for association or linkage with Alzheimer’s disease using family-based methodologies’, *Experimental Gerontology, 35:* 1353-1361.

Cooper, B. & Bickel, H. (1984), 'Population screening and the early detection of dementing disorders in old age: a review', *Psychological Medicine, 14:* 81-95.

Cooper, J.D., Salehi, A., Delcroix, J-D., Howe, C.L., Belichenko, P.V., Chua-Couzens, J.C., Kilbridge, J.F., Carlson, E.J., Epstein, C.J. & Mobley, W.C. (2001), ‘Failed retrograde transport of NGF in a mouse model of of down’s syndrome: Reversal of cholinergic neurodegenerative phenotypes following NGF infusion’, *Neurobiology, 98;* 18: 10439-10444.

Crawford, J.R., Gray, C.D. & Allan, K.M. (1995), ' The WAIS-R (UK): Basic psychometric properties in an adult UK sample', *British Journal of Clinical Psychology, 34:* 237-250.

Crayton, L. & Oliver, C. (1993), 'Assessment of Cognitive Functioning in Persons with Down's Syndrome who develop Alzheimer's Disease' in Berg, J., Karlinsky, H. & Holland, A. (eds), *Down's Syndrome and*

Alzheimer's Disease, Oxford: Oxford University Press.

Deb, S. & Braganza, J. (1999), 'Comparison of rating scales for the diagnosis of dementia in adults with Down's syndrome', *Journal of Intellectual Disability Research, 43 (Part 5):* 400-407.

De Groot, J.C., de Leeuw, F-E., Oudkerk, M., Hofman, A., Jolles, J. & Breteler, M.M.B. (2000), 'Cerebral white matter lesions and depressive symptoms in elderly adults', *Archives of General Psychiatry, 57:* 1071-1076.

Deng, W., Aimone, J.B. & Gage, F.H. (2010), 'New neurons and new memories: How does adult hippocampal neurogenesis affect learning and memory?' *Nature Reviews Neuroscience, 31 Mar,* doi: 10.1038/nrn2822, Accessed: 09.04.2010.

Dickson, D.W. (2001), 'Neuropathology of Alzheimer's disease and other dementias', *Clinics in Geriatric Medicine, 17;* 2: 209-228.

Dickson, D.W., Crystal, H.A. & Bevona, C. (1995), 'Correlations of synaptic and pathological markers with cognition of the elderly', *Neurobiology & Aging, 16:* 285.

Evenhuis, H.M. (1992a), *'Provisional Manual of the Dementia Questionnaire for Mentally Retarded Persons (DMR)'*, Zwammerdam: Hooge Burch.

Evenhuis, H.M. (1992b), 'Evaluation of a screening instrument for dementia in ageing mentally retarded persons', *Journal of Intellectual Disability Research, 36:* 337-347.

Evenhuis, H.M. (1996), ‘Further evaluation of the Dementia Questionnaire for Persons with Mental Retardation (DMR)’, *Journal of Intellectual Disability Research, 40;* 4: 369-373.

Evenhuis, H.M. (1997), ‘The natural history of dementia in ageing people with intellectual disability’, *Intellectual Disability Research, 41(Pt 1):* 92-96.

Furster, J.M. (1993), 'Frontal lobes', *Current Opinions in Neurobiology, 3:* 160-165.

Given, C.W., Given, B., Stommel, M., Collins, C., Ing, S. Franklin, S. (2007), ‘The caregiver reaction assessment (CRA) for caregivers to persons with chronic physical and mental impairments’, *Research in Nursing & Health, 15;* 4: 271-283.

Golding, E. (1989), *‘The Middlesex Elderly Assessment of Mental State’*, Bury St Edmunds: Thames Valley Test Company.

Greene, J.D.W., Hodges, J.R. & Baddeley, A.D. (1995), ‘Autobiographical memory and executive function in early dementia of Alzheimer type’, *Neuropsychologia, 33;* 12: 1647-1670.

Greene, J. & d'Oliveira, M. (1982), *'Cognitive Psychology. Methodology Handbook. Part 2: Parametric Designs and Tests'*, Milton Keynes: The Open University Press.

Grober, E., Dickson, D. & Sliwinski, M.J. (1999), 'Memory and mental status correlates of modified Braak staging', *Neurobiology & Aging, 20:* 573.

Janicki, M.P., Heller, T., Deltzer, G.B. & Hogg, J. (2007), 'Practice guidelines for the clinical assessment and care management of Alzheimer's disease and other dementias among adults with intellectual disability', *Journal of Intellectual Disability, 40;* 4: 374-382.

Hattori, M. Fujiyama, A., Taylor, T.D., Watanabe, H., Yada, T. & Park, H.S. (2000), 'The DNA sequence of human chromosome 21', *Nature, 405:* 311-319.

Hulme, C., Lee, G. & Brown, G.D.A. (1993), 'Short-term memory impairments in Alzheimer-type dementia: evidence for separable impairments of articulatory rehearsal and long-term memory', *Neuropsychologia, 31:* 161-172.

Kawas, C.H. & Katzman, R. (1999), 'Epidemiology of Dementia and Alzheimer Disease' in Terry, R.D., Katzman, R., Bick, K.L., *et al.*

(eds), *AlzheimerDisease Edition 2,* Lippincott, Williams & Wilkins, Philadelphia, 95.

Kay, B. (1993), 'Keeping it in the family', *Nursing Times, 89;* 22: 64-65.

Kearns, N.P., Cruikshank, C.A., McGuigan, K. & Riley, S.A. (1982), 'A comparison of depression rating scales', *British Journal of Psychiatry, 141:* 45-49.

Kendrick, D. (1985), *'Kendrick Cognitive Tests for the Elderly. Manual'*, Windsor: NFER-NELSON.

Kengen, M.M.F., Eurlings, H.A.L., Evenhuis, H.M. & de Boer, J. (1987), 'De Dementie Vragenlijst voor Zwakzinnigen (DVZ): Een onderzoeksinstrument voor de diagnostiek van seniele dementie bij zwakzinnigen', ['Dementia Questionnaire for Mentally Retarded Persons (DMR): An instrument for the diagnosis of senile dementia in mentally retarded persons'], *Ruit, 43:* 24-30.

Kennedy, A. & Neville, J. (1957), 'Sudden loss of memory', *British Medical Journal, 2:* 428-433.

Kolata, G. (1985), 'Down's syndrome - Alzheimer's linked', *Science, 230:* 1152-1153.

Kopelman, M.D., Wilson, B.A. & Baddeley, A.D. (1990), *'The Autobiographical Memory Interview'*, Bury St Edmunds: Thames

Valley Test Co.

Kramer, N.A. & Jarvik, L. (1979), 'Assessment of Intellectual Changes in the Elderly' in Raskin, A. & Jarvik, L. (eds), *Psychiatric Symptoms and Cognitive Loss in the Elderly*, Washington D.C.: Hemisphere.

Krishnan, K.R. (1991), 'Organic bases of depression in the elderlt', *Annual Review in Medicine, 42:* 261-266.

Krishnan, K.R., Hays, J.C. & Blazer, D.G. (1997), 'MRI-defined vascular depression', *American Journal of Psychiarty, 154:* 497-501.

Krishnan, K.R. & McDonald, W.M. (1995), 'Arteriosclerotic depression', *Medical Hypotheses, 44:* 111-115.

Lamberty, G.J. & Bieliauskas, L.A. (1993), 'Distinguishing between depression and dementia in the elderly: a review of neuropsychological findings', *Archives of Clinical Neuropsychology, 8:* 149-170.

Lishman, W.A. (1987), *'Organic Psychiatry: the Psychological Consequences of Cerebral Disorder'*, 2nd ed, Oxford: Blackwell.

Litvan, I., Grimes, D.A. & Lang, A.E. (2000), 'Phenotypes and Prognosis: Clinicopathologic Studies of Corticobasal Degeneration' in Litvan, I., Goetz, C.G., Lang, A.E., *et al.* (eds), *Corticobasal degeneration and Related Disorders (Advances in Neurology Volume 82),*

Lippincott-Raven, New York, 183.

Maclean, W.E., Ellis, D.N., Galbreath, H.N. & Halpern, L.F. (1991), 'Rhythmic motor behaviour of preambulatory motor impaired, Down syndrome and nondisabled children - A comparative analysis', *Journal of Abnormal Child Psychology, 19:* 319-330.

Markesberry, W.R. (1998), 'Vascular Dementia' in Markesberry, W.R. (ed), *Neuropathology of Dementing Disorders,* Arnold, New York, 293.

Milne, A. (2010), 'Dementia screening and early diagnosis: The case for and against', *Health, Risk & Society, 12;* 1: 65-76.

Morris, R.G. (1994a), 'Working memory in Alzheimer-type dementia', *Neuropsychology, 8;* 4: 544-554.

Morris, R.G. (ed) (1996), *'The Cognitive Neuropsychology of Alzheimer-Type Dementia',* Oxford, Oxford University Press.

Myers, A.J. & Goate, A.M. (2001), 'The genetics of late-onset Alzheimer's disease', *Current Opinion in Neurology, 14;* 433-440.

Myers, A., Holmans, P. & Marshall, H. (2000), 'Susceptibility locus for Alzheimer's disease on chromosome 10', *Science, 290:* 2304-2305.

Nash, J.M. (1997), 'Medicine', Time, March 24; 47-8.

O'Brien, J., Desmond, P., Ames, D. & Schweitzer, I. (1996), 'A magnetic

resonance imaging study of white matter lesions in depression and Alzheimer's disease', *British Journal of Psyhiatry, 168:* 477-485.

Powell, T., Brooker, D.J.R. & Papadopolous, A. (1993), 'Test-retest reliability of the Middlesex Assessment of Mental State (MEAMS): A preliminary investigation in people with probable dementia', *British Journal of Clinical Psychology, 32:* 224-226.

Pericak-Vance, M.A., Grubber, J. & Bailey, L.R. (2000), 'Identification of novel genes in late-onset Alzheimer's disease', *Experimental Gerontology, 35:* 1343-1352.

Pulsifer, M.B. (1996), 'The neuropsychology of mental retardation', *Journal of the International Neuropsychological Society, 2 (2):* 159176.

Rao, R. (1998), 'Depression after transient ischaemic attack: A clinically distinct subtype of vascular depression ?' *Archives of general Psychiatry, 55:* 753-754.

Raven, J.C., Court, J.H. & Raven, J. (1990), *'Raven's Progressive Matrices and Vocabulary Scales. Section 2: Coloured Progressive Matrices'*, Oxford: Oxford Psychologists Press.

Rhodin, J.A. & Thomas, T. (2001), 'A vascular connection to Alzheimer's disease', *Microcirculation, 8;* 4: 207-220.

Rossor, M.N. (1999), ‘Differential diagnosis of frontotemporal dementia: Pick’s disease’, *Dementia & Geriatric Cognitive Disorders, 10 (Suppl):* 43.

Ruitenberg, A., Kalmijn, S., de Ridder, M.A. (2001), ‘Prognosis of Alzheimer’s disease: The Rotterdam study’, *Neuroepidemiology, 20;* 3: 188-195.

Ruitenberg, A., Ott, A., van Swieten, J.C. (2001), ‘Incidence of dementia: Does gender make a difference?’ *Neurobiology of Aging, 22:* 575-580.

Schupf, N., Kapell, D., Nightingale, B., Lee, J.H., Mohlenhoff, J., Bewley, S., Ottman, R. & Mayeux, R. (2001), ‘Specificity of the fivefold increase in AD in mothers of adults with Down’s syndrome’, *Neurology, 57:* 979-984.

Scott, W.K., Grubber, J.M. & Conneally, P.M. (2000), ‘Fine mapping of the chromosome 12 late-onset Alzheimer disease locus: Potential genetic and phenotypic heterogeneity’, *American Journal of Human Genetics, 66:* 922-932.

Shallice, T. (1988), *‘From Neuropsychology to Mental Structure’*, New York: Cambridge University Press.

Shapiro, M.B., Haxby, J.V., Grady, C.L. (1992), 'The nature of mental

retardation and dementia in Down's syndrome: Study with PET, CT and neuropsychology', *Neurobiology of Ageing, 13:* 723-724.

Thompson, S.B.N. (1993b), *'Eating Disorders: A Guide for Health Professionals'*, London: Chapman & Hall.

Thompson, S.B.N. (1997), '*Dementia: A Guide for Health Care Professionals'*. Aldershot: Arena.

Thompson, S.B.N. (1999), 'Examining dementia in Down's syndrome (DS): Decline in social abilities in DS compared with ither learning disabilities', *Clinical Gerontologist, 20;* 3: 23-44.

Thompson, S.B.N. (2000a), 'The Central Executive System in people with Down's syndrome and dementia', *Clinical Gerontologist, 21;* 3: 3-32.

Thompson, S.B.N. (2000b), 'Investigation into Down's syndrome and dementia', *Journal of the Association of Practitioners in Learning Disability, 17;* 3: 10-14.

Thompson, S.B.N. (2002), 'Rate of decline in social and cognitive abilities in dementing individuals with Down's syndrome and other learning disabilities', *Under journal review.*

Thompson, S.B.N. (2009), Testamentary capacity and cognitive rehabilitation: Implications for head injure and neurologically

impaired individuals. *The Journal of Cognitive Rehabilitation, 27 (Fall):* 11-13.

Thompson, S.B.N. & Berry, A. (1997), 'The role of the clinical neuropsychologist', *British Journal of Therapy & Rehabilitation, 4;* 11: 584-588.

Thompson, S.B.N. & Berry, A. (1998), 'Neuroimaging and neuropsychology in the cognitive rehabilitation of stroke and head injury', *The Journal of Cognitive Rehabilitation, 16;* 3: 6-11.

van Belle, G., Uhlmann, R.F., Hughes, J.P. & Larson. E.B. (1990), 'Reliability of estimates of change in mental status test performance in senile dementia of the Alzheimer type', *Journal of Clinical Epidemiology, 43;* 6: 589-595.

Velayudhan, L., Poppe, M., Archer, N., Proitsi, P., Brown, R.C. & Lovestone, S. (2010), 'Risk of developing dementia in people with diabetes and mild cognitive impairment', *The British Journal of Psychiatry, 196:* 36-40.

Vincent, L.C. (1996), 'Neuropsychological assessment of developmental learning disabilities in adults', *Seminars in Speech & Language, 17 (3):* 183-200.

Vodegel-Matzen, L.B.L., van der Molen, M.W. & Dudink, A.C.M.

(1994), Error analysis of Raven test performance, *Personality & Individual Differences, 16;* 3: 433-445.

Wilson, B.A., Alderman, N., Burgess, P., Emslie, H. & Evans, J.J. (1996), *'Behavioural Assessment of the Dysexecutive Syndrome Manual'*, Bury St Edmunds: Thames Valley Test Co.

Wright, N. & Lindesay, J. (1995), A survey of memory clinics in the British Isles. *International Journal of Geriatric Psychiatry, 10:* 379-385.

Yapa, P. & Roy, A. (1990), 'Depressive illness and mental handicap: Two case reports', *Mental Handicap, 18;* 1: 19-21.

Young, E.C. & Kramer, B.M. (1991), 'Characteristics of age-related language decline in adults with Down's syndrome', Mental Retardation, 29; 2: 75-79.

APPENDICES

APPENDIX A

A NOTE ON THE MECHANISM OF THE ACETYLCHOLINE ESTERASE INHIBITORS (ACIs)

Since the late 1990s, there have been three prescribed "anti-dementia" drugs available to people with memory problems and suspected dementia. These were rolled out clinically, firstly for people with Alzheimer's disease though they were not widely available throughout the United Kingdom due to costs and budgets held within individual Primary Care NHS Trusts (PCTs) at that time. These drugs act on specific cholinergic receptor sites in the brain (the nicogenic and the muscarinic receptors) known to be associated with acetylcholine, the neurotransmitter linked to memory functioning.

The first medication to become more widely available was Aricept (Donepezil Hydrochloride, E2020) manufactured by Pfizer Pharmaceuticals. This had few side-effects except for reported cases of nausea, although some reports showed "Aricept-induced nightmares" . It was given in a single tablet dose. The second medication was Reminyl (Galantamine Hydrobromide) manufactured by Jansen-Cilag

Pharmaceuticals which was given in two doses (one dose, twice a day) and initially had fewer reported side-effects and evidence that it abated some of the behaviour disturbances sometimes seen with Alzheimer's disease, such as agitation. However, early on, there were reports that it should be avoided in cases of cardiac arrhythmias and heart disease. Towards the late 2000, increasingly there were reports that this medication is contra-indicated in the presence of heart problems and also the twice-daily dose is sometimes forgotten when it is self-prescribed by the patient or carer.

Exelon (Rivastigmine Tartrate) manufactured by Novartis Pharmaceuticals was found early on to have more side-effects than either of the other two choices of medication and is no longer prescribed widely. A newer medication, Ebixa (Memantine) manufactured by Lundbeck Pharmaceuticals acts on the glutaminergic receptor site in the brain and has been found to have fewer side-effects than all of the other drugs so far. Early clinical trials have shown that it may also have positive effects on cognitive functioning.

There remains insufficient data to conclude the benefits of any of these drugs in the treatment of vascular dementia; and to date, there is no

data on the treatment of either Alzheimer's disease or vascular dementia using any of these drugs for people with Down's syndrome and dementia. Clearly, there is scope for more focused research, particularly in the area of learning disabilities and dementia.

APPENDIX B

CONFIRMATION OF REGISTRATION AND RECOMMENDATION

-----Original Message-----
From: Fiona Knight
Sent: 09 March 2010 11:36
To: Naomi Bailey
Cc: Mark Hadfield
Subject: RE: PhD by Publication

Naomi -

The Graduate School is happy to approve Simon's prima facie case and he may now be registered onto the programme of PhD by Publication - with a maximum registration of 12 months.

Regards,
Fiona

The Graduate School

Bournemouth University

Professor John Fletcher
Head of The Graduate School

Ref: PGRPT/4303523/Thompson

Date 09 March 2010

To Whom It May Concern:

Student Number	4303523
Student Name	Simon Thompson
Start Date	01/03/2010
Mode of Study	Part Time
Duration of Study	12 Months
Programme End Date	28/02/2011

Please accept this letter as confirmation that the Graduate School has approved the above student's *prima facie* case and he may now be registered onto the programme of PhD by Publication at Bournemouth University with a maximum registration period of 12 months.

The student commenced this programme on 01/03/2010. Subject to successful completion within the normal timescales, it is expected that the programme end date will be 28/02/2011 and graduation is expected to be in November 2011.

Yours faithfully,

Professor John Fletcher

Head of The Graduate School
Bournemouth University

Poole House
Talbot Campus, Fern Barrow
Poole, Dorset BH12 5BB
United Kingdom

Tel +44 (0) 1202 965902
graduateschool@bournemouth.ac.uk
www.bournemouth.ac.uk

VAT Reg. No. GB 504 4921 66

Clinical Psychology Department,
Behavioural Resource Service,
315 Coxford Road,
Lordswood, Southampton,
Hampshire.
SO16 5LH

Dr Fiona Knight,
Graduate School Manager, Graduate School,
Bournemouth University, Talbot Campus,
Fern Barrow, Poole,
Dorset
BH12 5BB

30.4.2010

Dear Dr Knight,

RE: Advancing Knowledge into the Clinical Assessment of Dementia by Simon B. N. Thompson (PhD by Publication)

Due to my knowledge and experience in this area, I was asked to read Simon Thompson's comprehensive draft PhD thesis on dementia (which also covers learning disabilities).

From a content viewpoint, I would like to recommend his thesis for the consideration of an appointed External Examiner.

Yours sincerely,

Dr Sarah Hardy, BA (Hons), MSc, DClinPsy.
Principal Clinical Psychologist

APPENDIX C

COMPLETE LIST OF PUBLICATIONS IN TOPIC AREA

Books (9)

Thompson, S.B.N., ed, In Progress. *Clinical Psychology Handbook for Trainees: Theory & Practice Case Histories.*

Thompson, S.B.N., 2009; 2010. *Memory Matters.* Portsmouth: Oxford Press.

Thompson, S.B.N., 2006. *Dementia and Memory: A Handbook for Students and Professionals.* Aldershot: Ashgate.

Thompson, S.B.N., 2001; 2003. *Memory Problems: A Self-Help Guide for Patients, Carers, Health Professionals and Students in Training.* Chichester: Oxford Press.

Thompson, S.B.N., 2000. *A Self-Help Guide for Managing Everyday Memory Problems.* Chichester: Oxford Press.

Thompson, S.B.N., 1998. *A Self-Help Guide for People with Everyday Memory Difficulties.* Bishop Auckland: Cambridge Press.

Thompson, S.B.N., 1997. *Dementia: A Guide for Health Care Professionals.* Aldershot: Arena.

Peer Reviewed Journals (19)

Thompson, S.B.N., 2010. The dawn of the yawn: Is yawning a warning? Linking neurological disorders. *Medical Hypotheses, doi:10.1016/j.mehy.2010.08.002.*

Thompson, S.B.N., 2009. Testamentary capacity and cognitive rehabilitation – Implications for head-injured and neurologically impaired individuals. *The Journal of Cognitive Rehabilitation, 27(Fall),* pp.11-13.

Thompson, S.B.N., Ennis, E., Coffin, T. & Farman, S., 2007. Design and evaluation of a computerised version of the Benton Visual Retention Test. *Computers in Human Behavior, 23,* pp.2383-2393.

Thompson, S.B.N., Coates, T., Chaâbane, F., Cherry, P., Collins, L., Pennicott, H., Watson, F. & Rogan, N., 2006. Interdisciplinary clinic for adults with early onset dementia in a mental health NHS trust. *Clinical Gerontologist, 29(4),* pp.99-104.

Thompson, S.B.N., 2004. Efficacy of a memory clinic for patients with memory problems. *Chinese Journal of Clinical Psychology, 12(4),* pp.417-419.

Thompson, S.B.N., 2003. Rate of decline in social and cognitive abilities in dementing individuals with Down's syndrome and other learning disabilities. *Clinical Gerontologist, 26(3/4),* pp.145-154.

Thompson, S.B.N., 2002. Memory decline, Alzheimer's disease and vascular dementia: The clinical picture. *The Journal of Cognitive Rehabilitation, 20(2),* pp.12-18.

Thompson, S.B.N., 2001. Assessing dementia in Down's syndrome: A comparison of five tests. *Clinical Gerontologist, 23(3/4),* pp.3-19.

Thompson, S.B.N., MacDonald, J. & Coates, T., 2001. Improving visual memory with Aricept (Donepezil Hydrochloride, E2020) in mild-to-moderate Alzheimer's disease. *Clinical Gerontologist, 24(1/2),* pp.55-73.

Thompson, S.B.N. & Watson, F., 2001. 'Hollywood Wives': Education and support for spouses of inpatients with dementia. *The Journal of Dementia Care, 9(2),* p.38.

Thompson, S.B.N., 2000. Investigation into Down's syndrome and dementia. *Journal of the Association of Practitioners in Learning Disability, 17(3),* pp.10-14.

Thompson, S.B.N., 2000. The Central Executive System in people with Down's syndrome and dementia. *Clinical Gerontologist, 21(3),* pp.3-32.

Thompson, S.B.N., 1999. Examining dementia in Down's syndrome (DS): Decline in social abilities in DS compared with other learning disabilities. *Clinical Gerontologist, 20(3),* pp.23-44.

Thompson, S.B.N., 1999. Assessing dementia in people with learning disabilities for cognitive rehabilitation. *The Journal of Cognitive Rehabilitation, 17(3),* pp.14-20.

Kapur, N., Scholey, K., Moore, E., Barker, S., Brice, J. & Thompson, S.B.N., 1996. Long-term retention deficits in two cases of disproportionate retrograde amnesia. *Journal of Cognitive Neuroscience, 8(5),* pp.416-434.

Kapur, N., Thompson, S.B.N., Cook, P., Lang, D. & Brice, J., 1996. Anterograde but not retrograde memory loss following combined mammillary body and medial thalamic lesions. *Neuropsychologia, 34(1),* pp.1-8.

Thompson, S.B.N., 1996. Practical ways of improving memory storage and retrieval problems in patients with head injuries. *British Journal of Occupational Therapy, 59(9),* pp.418-422.

Thompson, S.B.N., 1994. A neuropsychological test battery for identifying dementia in people with learning disabilities. British Journal of Developmental Disabilities, XL, 2(79), pp.135-142.

Thompson, S.B.N., 1993, Doubting dementia in Down's syndrome. *The Psychologist, 6(9),* pp.397-400.

Professional Magazines (14)

Thompson, S.B.N., 2003. How good is your memory? *Horizons, 7,* pp.40-41.

Thompson, S.B.N., 2000. Memories… *MSF Works, 13,* pp.22-23.

Thompson, S.B.N., 2000. Neuropsychology and dementia. *NHS Newsline, 67,* p.4.

Thompson, S.B.N., 2000. New research for Alzheimer's disease. *NHS Newsline, 66,* p.4.

Thompson, S.B.N., 2000. Educational & support group for patients' relatives. *NHS Newsline, 65,* p.6.

Thompson, S.B.N., 1999. Managing everyday memory problems. *Psychology Post, 18,* p.7.

Thompson, S.B.N., 1999. Reflections on memory. *Therapy Weekly, 25(30),* p.6.

Thompson, S.B.N., 1999. A guide to remember. *Psychology Post, 11,* p.14.

Thompson, S.B.N., 1998. Self help guide for memory problems. *GP Connection,* Oct, p.8.

Thompson, S.B.N., 1995. Storage problems. *Therapy Weekly, 21(40),* p.7.

Thompson, S.B.N., 1993. Down's syndrome and Alzheimer's disease. *Therapy Weekly, 19(21),* p.8.

Thompson, S.B.N., 1992. The proof of the pudding. *Therapy Weekly, 19(16),* p.7.

Thompson, S.B.N., 1992. Tips to trigger memory. *Therapy Weekly, 19(7),* p.7.

Thompson, S.B.N., 1991. Aide-mémoire. *Therapy Weekly, 18(20),* p.7.

APPENDIX D

PAPERS PRESENTED IN TOPIC AREA

2010 'Ethics and Testamentary Capacity in Dementia.' *8th International Qualitative Research Conference, Bournemouth University, Bournemouth, UK, 6-8 Sep. Le Bâillement conference web site entry: http://baillement.com/dossier/thompson_dawn.html*

2010 'The dawn of the yawn: Is yawning a warning? Linking neurological disorders.' *La Première Conférence Internationale sur le Bâillement, l'Hôpital de la Pitié-Salpêtrière, Paris, France, 24-25 Jun.*

2010 'Alzheimer's Disease and Stroke - New Advances.' *Psychology @ BU, Celebrating Science & Practice in Psychology, Bournemouth University, UK, 17 May.*

2009 'Testamentary Capacity in Dementia.' *Poster Presentation, 6th International Congress on Vascular Dementia, Centre Convencions Internacional, Barcelona, Spain, 19-22 Nov.*

2009 'Improving Visual Memory with Aricept.' *Poster Presentation, 6th International Congress on Vascular Dementia, Centre Convencions Internacional, Barcelona, Spain, 19-22 Nov.*

2001 'Improving Visual Memory Performance with Aricept.' *Annual Eisai-Pfizer Conference on Anti-Dementia Drugs, Hilton Avisford Hotel, Arundel, UK, 4 Jun.*

2000 'Visual memory and Aricept in Alzheimer's Disease.' *Poster Presentation, British Psychological Society Annual Conference, Scottish Exhibition & Conference Centre, Glasgow, UK, 28-31 Mar.*

2000 'Potential Cognitive Benefits of Aricept for Improving Memory Functioning in Alzheimer's Disease.' *Inaugural Visiting Professorial Lecture, University of Portsmouth, Portsmouth, UK, 5 Oct.*

APPENDIX E

AWARDS GAINED IN TOPIC AREA

2010 **[£129 118].** Alzheimer's Society. Principal Investigator. 'Improving memory functioning in people with early onset Alzheimer's disease.' (Bid submitted)

2009 – 2013 **[£ 48 000].** Bournemouth University. Director of Studies/First Supervisor (with Dr B Parris). *'Investigating memory and language difficulties in Alzheimer's disease patients.'*

2004 – 2005 **[£ 2 500].** University of Portsmouth. Principal Investigator (with Dr E Ennis, T Coffin, S Farman). *'Design and evaluate a computerised version of the Benton Visual Retention Test.'*

2004 – 2005 **[£ 30 000].** Dorothy Allen Legacy. Principal Investigator (with Multi-Disciplinary Team). *'Risk factors in attendee carers and families of Adult Cognitive Assessment and Treatment clinic for Alzheimer's Disease.'*

2003 – 2004 **[£ 2 000].** The Nuffield Foundation Small Grants Scheme. (with Prof N Kapur). *'Design a recognition memory task of famous faces for patients with retrograde amnesia.'*

2003 – 2004 **[£ 1 750].** University of Portsmouth. Principal Investigator (with Dr E Ennis, T Coffin). *'Investigate the potential use of a computerised version of the Benton Visual Retention Test.'*

2001 – 2002 **[£ 750].** Jansen-Cilag Pharmaceuticals UK Ltd. Principal Investigator. *'Revise a self-help and professionals' guide for people with memory problems.'*

1999 – 2001 **[£ 64 000].** Pfizer Pharmaceuticals UK. Principal Investigator (with Multi-Disciplinary Team). *'Potential treatment benefit of Aricept on visual memory deficits in Alzheimer's disease.'*

2000 – 2001 **[£ 600].** Pfizer Pharmaceuticals UK Ltd. Principal Investigator. *'Design a self-help and professionals' guide for people with memory problems.'*

1999 – 2000 **[£ 500].** Novartis Pharmaceuticals UK Ltd. Principal Investigator. *'Design a book of memory strategies to help patients with everyday memory problems.'*

1997 – 1998 **[£ 800].** South Durham NHS Trust. Principal Investigator. *'Design and write a memory book for patients with memory difficulties and severe head injuries.'*

APPENDIX F

SUMMARY OF EXPERIENCE IN TOPIC AREA

Simon B N Thompson, studied Mathematics, Physics and Chemistry 'A' levels at Bablake School, Coventry and graduated with Bachelor of Arts (Honours) in Psychology at the University of Plymouth in 1982. He worked as an occupational therapy and art therapy helper when he published his first paper on the results of his BA (Hons) dissertation and also on working in psychiatry. Studying for the Postgraduate Diploma in Information Systems, he graduated in 1984 at the University of Portsmouth.

From 1984 – 1987, he carried out ground breaking research in stroke prognosis using proprioceptive electromyography and biofeedback at the School of Information Science, University of Portsmouth and graduated with a PhD in 1988. His work involved the design and development of a new computer expert system for stroke prognosis based on a stochastic model. He pioneered the Thompson Digital Switch for use in occupational therapy for the rehabilitation of stroke patients and those with leg injuries, particulary from snow skiing accidents and following

surgery. This went into commercial production from 1988. During this period, he also worked as Consultant for the development of computer software and peripherals for the paramedical professions.

After working as a psychology technician, in 1989, he trained in cognitive therapy with Professor Ivy-Marie Blackburn and also with people with eating disorders under Professor Chris Fairburn at the Cullen Clinic, Edinburgh, Scotland. He published his first book *Occupational Therapy for Stroke Rehabilitation (1990)* with Maryanne Morgan which is adopted as a core text by occupational therapists in training, and graduated from the Royal Edinburgh University Hospital in 1991 with MPhil in Clinical Psychology.

From 1991, he developed the clinical psychology and neuropsychology (dementia) service for people with learning disabilities covering North East Fife, Scotland and was later promoted in recognition of developing a new multiple disability service in Southampton. In 1993, he joined Professor Narinder Kapur at the Wessex Neurological Centre, Southampton University Hospital in clinical neuropsychological research, and published his second book *Eating Disorders: A Guide for Health Professionals*. In the following year, he became Associate Fellow of the British Psychological Society holding office between 1994 – 1997 as

Associate Editor of the British Psychological Society's *The Psychologist*, and Panel Member of the Policy Committee.

In 1997, he was appointed Assistant Director of the Teesside Clinical Psychology Training Programme and Consultant (Head) Clinical Neuropsychologist for South Durham. He published his third book *Dementia: A Guide for Health Care Professionals* followed by *A Self-Help Guide for People with Everyday Memory Difficulties* in 1998.

In 2000, he published *A Self-Help Guide for Managing Everyday Memory Problems* and was conferred Visiting Professor of Clinical Neuropsychology from the University of Portsmouth as one of the youngest people to be conferred.

In 2001, his sixth book *Memory Problems: A Self-Help Guide for Patients, Carers, Health Professionals and Students in Training* was published and also a landmark paper with Professor John MacDonald and Dr Tony Coates *Imroving visual memory with Aricept (Donepezil Hydrochloride, E2020) in mild-to-moderate Azheimer's disease* in *Clinical Gerontologist* which provided evidence of the first specific improvements in memory functioning using Aricept in short intervention.

Awarded £64k in research funding by Pfizer Pharmaceuticals, he investigated the cognitive benefits of Aricept followed by the efficacy of

a newly established assessment clinic for people with suspected dementia using £30k funding from a hospital legacy and in 2003, as part of the dementia care team, he received the prestigious 'NHS Team of the Year Award' for services to patients with dementia.

In 2004, as Expert Witness in Clinical Neuropsychology, he provided opinion on numerous criminal and civil cases at the Royal Courts of Justice, London as well as in the Combined Courts. He was appointed Principal Investigator (UK), EduPark European Consortium, conducting new research into Parkinson's disease in a seven-country European Union funded project attracting 1 million euros in funding.

In 2006, he published *Dementia and Memory: A Handbook for Students and Professionals* and in 2008 was appointed Senior Lecturer in Clinical Psychology and Neuropsychology at Bournemouth University. In the same year, *Neuroanatomy Made Easy* was published and in 2009, he was promoted to Programme Leader of the new Clinical Programme. *Memory Matters* was also published in the same year.

In 2011, *Neuroanatomy Made Really Easy* will be published together with a new core text for the MSc Foundations of Clinical Psychology *Clinical Psychology Handbook for Trainees: Theory & Practice Case Histories.*

He has held honorary positions: External Examiner, PhD Supervisor, External Clinical Assessor and has talked at a number of international conferences, BBC Radio and on film.