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Alzheimers Disease: Comprehensive Review of Aetiology, Diagnosis, Assessment Recommendations and Treatment.

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Abstract

Alzheimer's disease is a growing concern amongst clinicians and researchers, particularly because of the increase in referrals to hospitals and clinics. Longevity brings with it an increase in people with both organic and psychogenic disorders. The link between Down's syndorm and Alzheimer's disease helps our understanding of the disease but also presents us with complexity in terms of assessment and service provision.

Our understanding of the aetiology Alzheimer's disease has advanced; in is timely to consider how clinical assessment may also be improved.

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.

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).

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).

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 (Illustration 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. 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 (Illustration 2).

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.

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'.

Discussion

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.

Neuropathology and clinical signs of 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 peptide. At high concentrations vesicular amyloid beta 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).

Acetylcholine esterase inhibitors (ACI)

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 in the UK 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.

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.

Heredity

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 beta-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 e4 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).

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).

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.

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.

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 (Illustration 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 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).

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).

1. 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.

2. 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^{E} (gamma-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).

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).

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 (Illustration 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 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).

1. 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.

2. 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).

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 beta-amyloid precursor protein (beta-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: e2, e3 and e4 (Myers & Goate, 2001). In Caucasian populations, individuals who carry the e4 allele are three (heterozygotes) to eight (homozygotes) times more likely to develop Alzheimer's disease than individuals who do not harbour the e4 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.

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.

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.

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).

Recommendations for clinical assessment

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.

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. There have been several revisions in the past two decades but the WAIS-R continues to be used an recognised in the UK, especially in the courtroom for Expert Witness testimony and because of the provision to "pro-rate" items I order to shorten the overall administration duration of the test.

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. In 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 SEM 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 SEM) 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 SEM) 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 SEMs 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 SEMs. For the three WAIS-R IQs, average SEMs are 2.53, 2.74 and 4.14 IQ points for fullscale, verbal and performance, respectively.

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 Ab1-12, then set B1-12). Norms are only available for subjects aged 55 - 85 years (Raven, Court & Raven, 1995; p 63).

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.

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.

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.

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’.

Conclusion

The use of any measure for the clinical assessment of dementia, whether in people wih learning disabilities or in the ‘normal’ population carries with it limitations. Informed knowledge of these limitations allows use scientific choices which enable us to tailor our neuropsychological battery or adopt alternative measures.

Ultimately, there may be a compromise because of these limitations; however, scientific understanding has given us a better picture of the course of dementia than ever before. With the advancement of technology, such as MRI and fMRI, and PET and SPET scans, used in conjunction with neuropsychological tests administered at key time points including follow-ups, the clinician is better placed to make a more reliable diagnosis an prognosis than in the past. It is hope that this will also enlighten service providers in widening access to people with learning disabilities who also have dementia.

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Illustrations

Illustration 1

Differences between depression and dementia (adapted from Thompson, 1993, page 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

Illustration 2

Similarities and differences between dementia and other physical and psychological problems (adapted from Thompson, 1997, page 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; responsiveness slowness; non-responsiveness	Answers which usually accurate 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

Illustration 3

Adapted from Baddeley's (1992) Working Memory Model

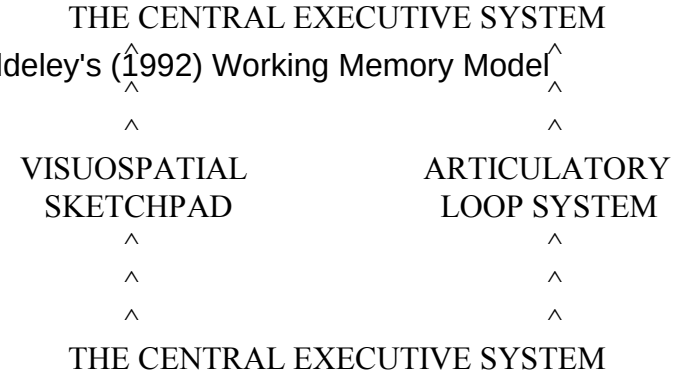

Illustration 4

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

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