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Division of Clinical Psychology
Faculty for Learning Disabilities

Dementia and People with Learning Disabilities

Guidance on the assessment, diagnosis, treatment and support of people with learning disabilities who develop dementia

CR155

September 2009

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Foreword

I am delighted to have been asked to write the foreword for this extremely important publication.

At the Down's Syndrome Association we have always been concerned about the need to make sure that everyone with Down's syndrome who develops dementia is able to gain access to proper assessment and treatment from the onset of their condition. There is now, generally, a better understanding about the particular link between Down's syndrome and dementia. That being said, there is still a great deal of confusion and ignorance about the treatment and care of those with learning disabilities who have dementia resulting in some quite inappropriate practice and provision.

The publication of this guidance is very timely as it deals with the assessment, diagnosis, treatment and the very necessary support needed for people with learning disabilities with dementia. I particularly welcome the fact that the guidance draws on comprehensive evidence based research and it brings together in partnership the expertise of some of the most eminent professionals currently working in services dealing directly with people with learning disabilities and their families.

I hope that the guidance will ultimately influence the development of comprehensive services and practices that will reduce the numbers of people with learning disabilities currently being failed by the provision that is available to them. It should be used and implemented by everyone working with people with learning disabilities including health and social care professionals, service providers, commissioners, care staff and families.

Carol Boys
Chief Executive
Down's Syndrome Association

Purpose of this report

This report is the result of a joint working group of the Learning Disability Faculty of the British Psychological Society and the Royal College of Psychiatrists.

The main purpose of the report is to enable those working in clinical and social care services to improve the quality of life of people with learning disabilities who develop dementia, by providing guidance to inform assessment, diagnosis, treatment and support. The guidance is aimed at clinicians in learning disability and older peoples' mental health services and services for younger people with dementia.

The constraints of time and practicality have meant that it has not been possible to produce a report that addresses every aspect of dementia in people with learning disabilities. Our main focus has been to highlight the key factors that we consider are the elements of an excellent service, and to help those working in services evaluate how they might help the increasing numbers of people with learning disabilities who are developing dementia given improvements in life-expectancy.

It is with these principles in mind that this report has been produced and has the following aims:

- to bring together available and relevant evidence-based practice with a consensus of clinical opinion and experience.
- to provide a framework for good practice and for the development of multi-agency care pathways.
- to promote effective and timely assessment, diagnosis, and treatment for people with learning disabilities suspected or confirmed as having dementia and to ensure quality support to them and their staff and other carers.
- to provide guidance for service providers, developers and commissioners.
- to provide a set of standards of good practice against which service provision can be benchmarked and audited.
- to promote the development of comprehensive and effective local services and to reduce the number of individuals who are failed by current service provision.
- to provide a framework for training health and social care professionals and paid support staff and carers.
- to guide the future development of services.

The report was produced through the combined work of members of the working group and drew on the published evidence base and from the working group members' extensive clinical experience in this area. Although this report has been undertaken primarily by psychologists and psychiatrists, we recognise that people with dementia must have effective multi-agency and multi-disciplinary services. We believe that this report will therefore be relevant to anyone who has an interest in dementia and people with learning disabilities, including health and social care professionals, families, paid staff, advocates, service providers and commissioners.

We have not been prescriptive within the document, particularly about the choice of assessment tools, because the evidence is not yet available to support particular instruments. Decisions will still need to be made locally depending on local resources and configurations. However, the report is intended to highlight the specific issues that people with learning disabilities and dementia present, and to ensure that local services are timely, effective and ensure that the person continues to have a high quality, safe and person-centred lifestyle as the dementia progresses.

This is a rapidly developing area with new knowledge and practice developing all the time. We envisage that these guidelines will need a regular review cycle to update them, as new evidence is published.

Finally, the report hopes to complement other publications and guidance in this area and to provide a way forward for supporting people with learning disabilities who develop dementia.

The audit tool at the end of the guidance is based on that developed by the Royal College of Psychiatrists, the British Psychological Society and Royal College of Speech and Language Therapists for their guidance on challenging behaviour – *Challenging Behaviour: A Unified Approach* (RCPsych et al., 2007). We are grateful for the permission of that group to adapt the tool for this guidance.

Section 1 – Context

This document is concerned with standards of clinical practice in assessing, diagnosing treating and supporting people with learning disabilities who develop dementia.

1.1 Dementia publications

Interest in the field of dementia in the non-disabled population has increased significantly over the past three years, with a range of publications focusing on the needs of both older people and those with dementia.

In 2005 the Department of Health and the Care Services Improvement Partnership (CSIP) published *Everybody's Business: Integrated mental health services for older adults: A service development guide* (CSIP, 2005). This document sets out the essentials for a service that works for older people's mental health in general. It addresses memory assessment services to enable the early diagnosis of dementia for all. It describes integrated community mental health teams whose role includes the management of people with dementia with complex behavioural and psychological symptoms. It recognises that effective services for older people with mental health problems and learning disabilities will only be achieved through joint working and efficient partnerships. Success requires staff and professionals in both older people's mental health and learning disability services to be prepared to do things differently.

The National Institute for Health and Clinical Excellence (NICE) and the Social Care Institute for Excellence (SCIE) published a joint clinical guideline on the management of dementia in 2006 (NICE/SCIE, 2006). Key recommendations included: integrated working across all agencies; provision of memory assessment services as a point of referral for diagnosis of dementia; assessment, support and treatment (where needed) for carers; assessment and treatment of non-cognitive symptoms and behaviour that challenges; dementia care training for all staff working with older people; and improvement of care for people with dementia in general hospitals.

Several reports, primarily concerned with dementia in the non-disabled population, have highlighted inadequacies in all services for people with dementia.

Raising the Standard (Royal College of Psychiatrists, 2006) concluded that there needs to be better co-ordination between agencies. Older people with mental health problems and learning disabilities may have complex needs and may need to draw on expertise from a number of areas including specialist mental health, older peoples' mental health, generic dementia, and learning disability services. It also advocated the development of joint working practices between learning disability, mental health and older people's services. In some places this may involve the formation of joint teams.

The National Audit Office in their report (NAO, 2007) was critical of the quality of care received by people with dementia and their families. It found that the size and availability of specialist community mental health teams was extremely variable and that the confidence of GPs in spotting the symptoms of dementia was poor and lower than it had been in 2000. They also commented on deficiencies in carer support. The report concluded that, overall, services are not currently delivering value for money to taxpayers or people with dementia and their families; that too few people are being diagnosed or being diagnosed early enough, and that early intervention is needed to improve quality of life; and, finally, that services in the community, care homes and at the end of life are not delivering consistently or cost-effectively against the objective of supporting people to live independently as long as possible in the place of their choosing. The NAO advocated a 'spend to save' approach, with upfront investment in services for early diagnosis and intervention and improved specialist services, community services and in general hospitals, resulting in long-term cost savings from prevention of transition into care homes and decreased length of hospital stay.

The Commission for Social Care Inspection reported on people's experiences of living in a care home in their report *See Me, Not Just The Dementia* (CSCI, 2008). Amongst its findings, the study demonstrated that the quality of staff communication with people with dementia has a major impact on their quality of life. This confirms the importance of a positive communication style with people with more advanced dementia. The findings support the importance of well-trained and supported staff working in homes committed to person-centred care. Care home managers need to provide leadership, home ethos, staff support and training to ensure excellent personalised care; and to develop ways of assessing the well-being of the people with dementia they are caring for. Local councils and Primary Care Trusts need to procure services at a price that allow for the one-to-one communication and trained staff essential to people's quality of life and well-being.

As a result of these documents and the increase in interest in dementia, the Government has made dementia a priority and consulted on a National Dementia Strategy (DH, 2008) prior to the publication of the final version *Living well with dementia: A National Dementia Strategy* (DH, 2009). The strategy has three key steps: improved awareness and understanding of dementia and removal of the stigma that surrounds it; early diagnosis and intervention; and improving the quality of care for people with dementia by developing a range of services for people with dementia and their carers which fully meets their changing needs over time. The strategy, which has 17 objectives, will form a five year plan relating to all people with dementia, including people with learning disabilities.

1.2 Assumptions made within the guidance document

Within this document, a number of assumptions are made throughout. These include the need for effective partnership working between all agencies involved in the care of people with learning disabilities and dementia as highlighted in reports cited in the previous section. The Working Party recognises that each area will be configured to meet local need, but effective care can only be provided when there is good partnership working within health services – between learning disabilities and older people's services, and across statutory, private and voluntary agencies.

However, regardless of how each service is configured, the working party has assumed that certain principles and ways of working are already integral to the delivery of services for people with learning disabilities, and that these will also be available to people with learning disabilities who develop dementia. It has been assumed that services already deliver care in line with *Valuing People* (DH, 2001) ensuring that everyone who wants one has a Person Centred Plan and a Health Action Plan, as well as an individualised care plan.

It has also been assumed that services will be delivered in line with both the *Mental Capacity Act* (Department for Constitutional Affairs, 2005) and the *Human Rights Act* (Department for Constitutional Affairs, 1998).

Section 2 – Epidemiology

2.1 Epidemiology of dementia in people with learning disabilities.

There have been significant improvements in the mean life-expectancy of people with learning disabilities from as little as an estimated 18.5 years in the 1930s to 59 years in 1970s to 66 years in the 1990s (Braddock, 1999). The life–expectancy of those with a more severe level of disability, however, remains reduced compared to the general population, as does the mean life-expectancy of people with Down's syndrome, which has been estimated at 55 years (Holland et al., 2000). The life-expectancy of those with mild learning disabilities now approaches that in the general population of a similar socio-economic status. Given these improvements, it has been predicted that the proportion of people with learning disabilities over 65 years of age will have doubled by 2020 (Janicki & Dalton, 2000) and that over a third of all people with learning disabilities will be over 50 years of age by that time (McConkey et al., 2006). It is because of these factors that consideration needs to be given to those age-related illnesses that most commonly occur in later life, such as dementia. Down's syndrome is the most frequent known cause of mild and severe intellectual disability (Minns, 1997) and may account for 15–20 per cent of the learning disabilities population (Pulsifier, 1996). Overall, neither the expected nor observed birth prevalence of Down's syndrome in England and Wales is declining (Huang et al., 1998), although Ekelund et al (2008) reported that a new national screening policy for Down's syndrome in Denmark halved the number of babies born with the condition between the years 2000 to 2006.

Estimating prevalence rates of dementia can be problematic because of a number of methodological issues that are broadly concerned with, first, the diagnosis of illnesses such as dementia in a population which has pre-existing cognitive and functional impairments and, second, the problems with respect to establishing a true population sample of people with learning disabilities. However, there is evidence from several studies that people with learning disabilities have an increased risk of developing dementia with increasing age compared to that observed in the general population. In particular, those with trisomy 21 resulting in Down's syndrome have an earlier age-related risk of developing dementia of the Alzheimer-type. There is very limited research investigating the specific risk of dementia and other age-related disorders in those with other specific syndromes, some of which are associated with a reduced life-expectancy.

2.2 Prevalence rates of dementia among older people with learning disabilities (excluding people with Down's syndrome)

The following studies have investigated rates of clinical dementia among people with learning disabilities living within the community. In each case, established or modified criteria based on systems for the diagnosis of dementia have been used. Moss & Patel (1995) reported that 12 per cent of a group of people with learning disabilities over age 50 years had dementia. Cooper (1997) found that the rate of dementia increased as expected in a population-based study with just over 20 per cent of those over the age of 65 years meeting criteria for dementia. Strydom et al. (2005) reported findings from a two stage population based survey of adults with learning disabilities (without Down's syndrome) across several London Boroughs. They found that prevalence rates varied depending on the diagnostic criteria used, with DSM-IV criteria resulting in the highest rate and ICD-10 the lowest. Rates for dementia, of whatever cause, using DSM-IV criteria were 13.1 per cent in those 60 years and over and 18.3 per cent in those 65 years or over. The four most common causes based on clinical observations were Alzheimer's disease, Lewy body dementia, vascular dementia, and fronto-temporal dementia. A more recent study (Strydom et al., 2007) showed that DSM–IV dementia criteria were more inclusive, with diagnosis using ICD–10 excluding people with even moderate dementia.

This compares to prevalence rates in the general population, quoted by the Eurodem Consortium of 1 per cent for 60- to 65-year-olds to 13 per cent for 80- to 85-year-olds and 32 per cent for 90 to 95-year-olds (Hofman et al., 1991). See Figure 1 below.

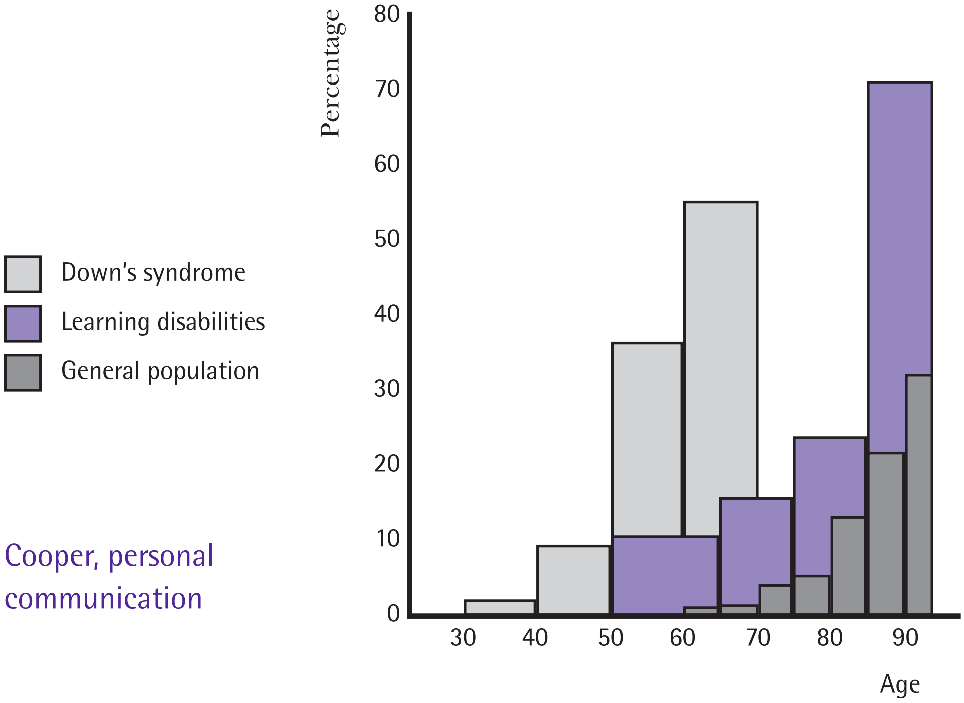
2.3 Prevalence rates of dementia among people with Down's syndrome

The association between Down's syndrome and the risk of 'precipitated senility' was first reported by Fraser & Mitchell in 1876, with Struwe (1929) describing the significant Alzheimer-like neuropathological changes in the brains of people with Down's syndrome.

Since then case reports, cross-sectional and longitudinal studies have all confirmed an increase in the prevalence rates of clinically diagnosed dementia with increasing age that starts when people with Down's syndrome are in their 30s and steadily increases in prevalence into the 60s. These rates are not as great as neuropathological studies initially suggested would be the case and, whilst the precise rates differ between studies, all agree that not all people with Down's syndrome in later life develop the pattern of memory loss and functional decline characteristic of dementia. Risk factors in understanding the development of dementia were reviewed by Bush and Beail (2004).

Studies have reported rates of dementia meeting the necessary criteria that start at a few percent between 30 and 39 years of age, increasing to between 10 per cent to 25 per cent in the 40- to 49-year-old group and to 20 per cent and 50 per cent in the 50- to 59-year-old group and between 30 per cent and 75 per cent aged 60 years or older (Hewitt et al., 1985; Wisniewski et al., 1985; Lai & Williams, 1989; Holland et al., 1998). It is generally agreed that the peak incidence is in the early fifties. A number of studies have reported that behavioural changes, rather than functional memory decline, are the early signs of developing dementia and Ball et al. (2006) have proposed that this may be explained by limited reserve capacity of the frontal lobes of people with Down's syndrome. Longitudinal neuropsychological studies have also found that some discrete cognitive abilities, as measured using established neuropsychological tests, show evidence of increasing impairment over time starting in the early and pre-clinical stages of dementia. Thus memory and orientation are affected early, with praxis, language and visuo-spatial skills becoming impaired later in the progression of the disorder (see Ball et al., 2006 for review).

Figure 1: Comparative rates of Dementia – Down's syndrome, Learning disabilities, General population



Cooper, personal communication

Figure 1 summarises the age-related prevalence rates of dementia in people with Down's syndrome, those with learning disabilities without Down's syndrome, and in the general population. The exact rates have to be considered with caution but the trend represented in this figure is now increasingly accepted. The early presentation and course of dementia is now well established for people with Down's syndrome. For those with learning disabilities but without Down's syndrome, age-related prevalence rates are brought forward to a small degree compared to the general population but not to the same extent as for people with Down's syndrome. This latter group would appear to have a uniquely early risk for developing dementia, almost invariably of the Alzheimer's-type. For the former group the full range of causes of dementia is observed.

Amyloid is the protein that, in an insoluble form, forms the neural plaques characteristic of Alzheimer's disease. The gene coding for the Amyloid Precursor Protein (APP) is located on chromosome 21, and this is likely to account for the increase in risk of Alzheimer's disease in people with Down's syndrome. Children with Down's syndrome have been found at post mortem to have evidence of diffuse cerebral amyloid deposition and in adult life plaques and tangles characteristic of Alzheimer's disease are found. The assumption therefore is that this slow deposition of amyloid in the brain leads to a cascade of adverse neural events over time and ultimately to the full pathology of Alzheimer's disease. However, whilst the brain pathology characteristic of Alzheimer's disease would seem to be universal in later life, it is clear that not all older people with Down's syndrome develop the clinical features of dementia. The reason for this remains unclear.

Key points:

- People with learning disabilities have a higher risk of developing dementia compared to the general population, with a significantly increased risk for people with Down's syndrome and at a much earlier age.
- Life expectancy of people with Down's syndrome has increased significantly.
- The incidence and prevalence of Down's syndrome is not decreasing.

Section 3 – Baseline and monitoring

3.1 Baseline screening, reactive and prospective monitoring

Services will need to consider what type of service they should offer to people with learning disabilities who may develop dementia. Services will need to:

- establish a baseline for every adult with Down's syndrome whilst they are healthy;

and

- provide reactive screening, i.e. assessing for dementia in any adult with learning disabilities after concerns have been raised;

and they may consider

- undertaking prospective screening for dementia for adults with Down's syndrome conducted at intervals from the age of 40 or 50 onwards.

As long ago as 2001, good practice guidance from the Foundation for People with Learning Disabilities (Turk et al., 2001) recommended that every service for people with learning disabilities should set up a register of adults with Down's syndrome, conduct a baseline assessment of cognitive and adaptive functioning before the age of 30 years, develop specialist skills in this area, offer training to other professionals, front line staff and carers, and seek high quality co-ordination between agencies. Despite this, the availability of screening and treatment across the UK is inequitable. Although in most areas reactive assessment is provided for those with signs of deterioration, very few services offer baselines and prospective screening such as that described by McBrien et al. (2005) to all young adults with Down's syndrome,

Types of tools used for dementia screening are described in Section 6 – Assessment.

3.2 The importance of baseline screening

There are several reasons why it is important to establish an accurate baseline of cognitive and adaptive functioning for people with learning disabilities and particularly those with Down's syndrome (Bush & Beail, 2004).

1. There is no definitive 'test' for dementia. Its presence is a matter of eliciting a clinical history suggesting dementia and establishing, retrospectively or prospectively, evidence of change in function from a known baseline and then excluding other diagnoses that may mimic dementia. In the mainstream population it is much more straightforward to gauge pre-morbid functioning from self report, employment history, etc than it is in the population of people with learning disabilities where self-report is very limited and few paid carers are in possession of a full history. Unless a baseline is established when the person is healthy, it is very difficult to establish whether there has been a deterioration later in life. By the time an individual is referred with concerns, considerable deterioration may have occurred and an accurate account of pre-morbid function may be difficult to construct. Longitudinal data is then needed to establish decline from a baseline as it is not possible to

compare results of an assessment with an 'average' result for the Down's syndrome, learning disabilities or general population.

2. Early detection of dementia relies on a good baseline. Prompt diagnosis ensures that attention can be paid in a timely way to necessary changes to a care package, medication, preparing family carers and support staff for the inevitable changes and challenges that dementia will bring. This may help the person to access cognitive enhancers in line with NICE eligibility guidelines.
3. Turk et al. (2001) recommended that baseline be established by the age of 30. Carr (2000) demonstrated stability in intellectual ability and daily living skills for her cohort of people with Down's syndrome over the age period 21 to 30 years. This suggests that a baseline conducted in the 20s would capture people post-maturity and prior to any cognitive decline.

3.3 Reactive monitoring

Reactive monitoring is the most common provision made by learning disabilities services. However, the reliability and efficiency of reactive screening can be greatly enhanced if a baseline assessment is available.

Reactive screening means conducting a dementia assessment after concerns about deterioration have been raised. This relies on carers noticing relevant signs of change and making a referral to the GP or learning disability service. This does not always happen. Due to high turnover of staff in many learning disabilities residential settings, staff may not be aware of changes in a person's presentation. To enhance this, checklists can be used at annual reviews (e.g. Whitwham et al., 2007), and staff awareness training can be provided for residential homes.

3.4 Prospective monitoring

Prospective monitoring entails a regular assessment to check for early signs of dementia by repeating the baseline assessment. This necessitates, as do baseline assessments, having a register of all adults with Down's syndrome and additionally a method of recalling people for a re-screen at set intervals.

The frequency of prospective screening for dementia should be matched to the rising risk with age. For example, first baseline screen might take place at 30 years; then every two years for those in their 40s; and annually for those aged 50 and over. Screening is non-invasive and usually enjoyable for the participant. Nevertheless, it needs to be justified in terms of likely gain, and there needs to be full consideration of the issue of informed consent. Some services may decide against it for reasons of intrusion, commissioning intentions and/or resource limitations. Apparent changes in scores on cognitive screening assessments do not in themselves indicate dementia but do require further investigation.

One argument in favour of prospective screening concerns the known health risks for all people with Down's syndrome. Many treatable illnesses can produce symptoms of cognitive decline independent of dementia. The evidence is that all too often, such disorders remain undetected in learning disabilities populations (Band, 1998). This applies particularly to people with Down's syndrome all of whom are prone to certain health problems (as described in Section 4). Many carers and indeed some GPs are not aware, for example, of the need for routine thyroid function tests for adults with Down's syndrome. There are useful factsheets, available free of charge on the internet from the Down's Syndrome Association (Dennis & Marder, 2006), that can be used to raise awareness of this issue with carers and doctors.

For example, an audit of six months of prospective screens carried out in one learning disability service showed that of 66 screenings of apparently healthy adults with Down's syndrome, 12 (18 per cent) revealed concerns that had not previously been identified by carers. The majority were about physical or mental ill health that could be immediately treated, whilst three required action by social services, including one requiring the protection of the Vulnerable Adult protocol (McBrien, personal communication).

The evidence does not currently indicate a need for prospective screening for people with learning disabilities without Down's syndrome. Screening for this group should be in response to concerns raised about changes in memory, behaviour or mood.

Key points:

- It is advisable to assess every adult with Down's syndrome at the age of 30 to establish a baseline against which to compare future suspected changes in functioning.
- It is worth considering screening all adults with Down's syndrome over 40 regularly because of the increased risk of dementia and the prevalence of undetected treatable illnesses.

Section 4 – Possible reasons for apparent decline in functioning

Commonly referrals are not clear in specifying the type of decline, but are often presented as behaviour changes and/or changes in functional abilities. It is important to remember that some changes may be part of the normal ageing process. The list below, whilst not exhaustive, describes the most common reasons for change in ability. It is important to recognise that two or more conditions can co-exist.

1. **Physical problems** include such conditions as hypothyroidism, anaemia, uncontrolled epilepsy, chronic infections, nutritional deficiencies. A thorough physical examination and relevant clinical tests are required at the time of initial assessment. These would need to be repeated from time to time as necessary.
2. **Sensory impairment:** People with learning disability in general and those with Down's syndrome in their middle/old age are likely to develop hearing and visual impairments. Cataract is the most likely cause of visual impairment and earwax the most likely cause of a conductive hearing impairment.
3. **Mental health problems:** The most common differential diagnosis is depressive illness (McBrien, 2003) but other conditions like the exacerbation of an existing psychotic disorder can mimic the presentation of dementia.
4. **Iatrogenic causes:** Medications with anticholinergic side effects can cause cognitive impairments in elderly people. Use of high dose psychotropic/anti-epileptic medications and multiple medications can contribute to cognitive impairment as well.
5. **Impact of life events:** People with learning disabilities in their middle age can face a number of life events such as loss of a parent or long-term carer, moving away from home, loss of day activities. In some individuals, the impact of life events may lead to a regressive state with apparent loss of skills.
6. **Acute Organic Brain syndrome:** This may co-exist with dementia or be part of the differential diagnosis.
7. **Abuse:** Current or recent physical, emotional or sexual abuse in people with learning disabilities may result in loss of skills and regression and the development or exacerbation of behaviour problems that might superficially mimic dementia.
8. **Impact of poor environment:** Unsuitable environment associated with lack of stimulation, isolation and lack of opportunities for positive interaction can lead to loss of skills.
9. **Dementia:** the typical presentation of dementia is one of gradual loss of skills along with change in personality and cognitive decline.

Key points:

- The common differential diagnoses for individuals with Down syndrome presenting with loss of skills are depressive illness, sensory impairments (hearing or visual), hypothyroidism and dementia.
- Sometimes more than one of these conditions can co-exist.

Section 5 – Clinical presentation of dementia

5.1 Criteria for dementia

Within the general population, diagnostic criteria have been developed in order to improve the accuracy of the clinical diagnosis of dementia (Prasher, 2005). These include the ICD-10 and DSM (IV) criteria that are summarised below.

ICD-10 (1992) criteria (summary)

1. Evidence of decline in memory, most evident in the learning of new information. The impairment applies to both verbal and non-verbal material and is sufficient to interfere with everyday function.
2. A decline in other cognitive abilities and daily living skills, characterised by deterioration in judgment and thinking such as planning and organising, and in the general processing of information, to a degree leading to impaired functioning in daily living. These include:
    - Language comprehension and expression.
    - Perception.
    - Praxis.
    - Executive function.
    - Usual daytime activities.
    - Use of household utensils and equipment.
3. Absence of clouding of consciousness/delirium.
4. Decline in emotional control, motivation or social behaviour in at least one of the following:
    - Emotional lability.
    - Irritability.
    - Apathy.
    - Coarsening of social behaviour.
5. The duration in changes in memory must be longer than 6 months.

DSM (IV) criteria (Morrison, 1995)

The development of multiple cognitive deficits such as manifested by impaired memory, long or short-term, can't learn new information or can't recall information previously learned and is distinguished by:

1. One (or more) of the following cognitive disturbances:
    - Aphasia (language disturbance).
    - Apraxia (impaired ability to carry out motor activities despite intact motor function).
    - Agnosia (failure to recognise or identify objects despite intact sensory function).

- Disturbance in executive functioning (i.e., planning, organising, sequencing, abstracting).

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

2. The decline in mental functioning begins gradually and worsens steadily.
3. The cognitive deficits above are not due to any of the following:
    - Other central nervous system conditions that cause progressive deficits in memory and cognition (e.g., cerebrovascular disease, Parkinson's disease, Huntington's disease, subdural hematoma, normal-pressure hydrocephalus, brain tumor).
    - Systemic conditions that are known to cause dementia (e.g. hypothyroidism, vitamin B-12 or folic acid deficiency, niacin deficiency, hypercalcemia, neurosyphilis, HIV infection).
    - Substance-induced conditions.
4. They aren't better explained by another Axis I disorder such as Depressive Disorder or Schizophrenia.

5.2 Course of the disease

True dementia (as opposed to pseudodementia due to depressive illness, or dementia like symptoms due to thyroid disorder) is at present untreatable and has a progressive course. In the non-learning disabled population each of the different types of dementia have characteristic clinical features. The presentation and course of dementia has been studied in people with Down's syndrome but less so in those with learning disabilities due to causes other than Down's syndrome. However, some general points can be made.

People with Down's syndrome

The presentation and course of dementia in people with Down's syndrome has been increasingly well established. The most common form of dementia in people with Down's syndrome is Alzheimer's disease. It is known that the brain pathology of Alzheimer's disease is almost universally found in later life in people with Down's syndrome. Vascular disease, and, therefore, the risk of dementia of a vascular origin, is rare.

Dementia in people with Down's syndrome may present atypically with changes in behaviour and/or personality that can precede the full clinical picture of dementia by some years. The same may also be true for other people with learning disabilities.

Dementia in people with Down's syndrome may be associated with the onset of seizures for the first time in that person's life.

The middle and later course of dementia (specifically Alzheimer's disease) in people with Down's syndrome and for those with other causes for their learning disabilities are similar in characteristics to those experienced by people in a similar stage of dementia but without pre-existing learning disabilities.

The time course of dementia in people with Down's syndrome with dementia has been reported to be more rapid than in the general population. Whether this is the case or not is uncertain as it may be a manifestation of the difficulties and delay in making an early diagnosis (Bush & Beail, 2004). By the time the diagnosis is made the dementia may already be advanced.

People with learning disabilities are at high risk for additional co-morbid illness that might impact on the way that dementia presents and its progression over time. Unexpected deterioration or changes in presentation or course should be investigated in case there are treatable additional physical or psychiatric co-morbid illnesses.

Atypical presentations in people with Down's syndrome

There have been anecdotal reports of people with Down's syndrome in their teens or early adult life who deteriorate, often following a life event, and either never or, only after many months or years, recover. This has not been reported in the literature but the characteristics of the decline may superficially resemble that of dementia or depressive illness but it neither seems to progress (as would be expected with dementia) or resolve (as would be expected with depressive illness).

The clinical picture is dominated by the development of a general slowness in mental and/or physical activity, apparent loss of interest in previous activities, and a level of functioning that is below that previously observed. At present it is unclear how such problems should be best conceptualised. If depressive illness is a possible factor, a trial of anti-depressant medication may be indicated, with careful monitoring of outcomes. Regular review of neuropsychological function is helpful in order to check that there is no progressive disorder such as dementia. Therapeutically the approach taken is primarily a rehabilitative one with attempts to help the person progressively back to their previous state of function.

People with learning disabilities without Down's syndrome

For those people with learning disabilities not due to Down's syndrome, the picture is less clear cut. There is more likely to be a range of pathologies resulting in dementia, as is the case in the general population, and for the same reason differences in presentation and the course of dementia.

Experience from the study of older people with Down's syndrome suggests that the clinical presentation may be affected by an interaction between how brain development is affected due to having Down's syndrome and the developing pathology of Alzheimer's disease. The same is likely to be true among people with learning disabilities not due to Down's syndrome as there will be a great variation across this population in terms of level of learning disabilities. The following are likely to be useful guiding principles with respect to the presentation and course of dementia in those with learning disabilities not due to Down's syndrome:

- The presentation and course of dementia among people with mild learning disabilities is likely to be similar to that which is observed in the general population.
- The presentation and course of dementia in people with more severe learning disabilities may initially be atypical and present with changes in behaviour and for this reason dementia may not be suspected. As the illness progresses careful questioning of staff and carers may identify evidence of developing memory and functional impairments.

- The age of onset of dementia in this group of people may be a few years earlier than is commonly found in the general population but not as early as in people with Down's syndrome.
- As with those in the general population it is important to investigate the likely cause of dementia as the full range of causes for dementia are likely to be found in this group of people and this may have important therapeutic and management implications.
- There is evidence that the onset of epilepsy in later life for the first time may be a marker for developing dementia in people with Down's syndrome. Whether this is the case for other people with learning disabilities is uncertain but onset of epilepsy in later life for the first time should always be investigated and increasing difficulty controlling pre-existing epilepsy may be an indication for considering the possibility of developing dementia.

Key Points:

- The course of dementia in people with Down's syndrome has been well studied, and the most likely cause is Alzheimer's disease.
- A small number of young people with Down's syndrome seem to present with decline in their teens or early twenties, often with no clear aetiology.
- People with learning disabilities without Down's syndrome who develop dementia may have the same range of pathologies as the general population.

Section 6 – Assessment

The process of assessment essentially has three stages that follow NICE/SCIE guidance on dementia (2006).

1. A systematic history from the person him/herself and from an informant who has known that person over time to establish the presentation and course of the presenting problem/s. Collateral information on the setting/s (home/day centre) is important. Changes in the individual may be due to wider systemic issues within the setting that the informant is either not aware of or not prepared to mention (e.g. Care Quality Commissio (CQC) concern, Safeguarding Adult investigations) or directly caused by concurrent abuse that is not recognized or not acted upon. This can be identified through informant interview and direct observation as well as good intelligence amongst members of a multi-disciplinary team.
2. Physical and mental state examinations, cognitive assessments and other investigations to enable the evaluation of present functioning and the identification of other possible causes of decline. The medical investigations should be guided by the clinical picture but invariably include investigations of a person's basic physical state (e.g. kidney and liver function and the presence or not of anaemia) and specific tests, such as measures of thyroid function, or specialist assessment of hearing and/or vision. Where the clinical picture is unusual or the diagnosis in doubt a CT or MRI brain scan may be indicated.
3. The evaluation of findings against known criteria for dementia and for other physical and mental disorders in order to arrive at a definitive diagnosis. For people with Down's syndrome, three particular disorders are most common and their presentation may mimic that of dementia as well as co-exist with dementia and thereby make the disabilities associated with the development of dementia significantly more pronounced. These are:
    - Depressive illness.
    - Under-active thyroid gland (hypothyroidism).
    - Visual and/or hearing impairments.

The results of all three parts of the diagnostic process need to be brought together to arrive at the formulation. Results of specific tests are always a guide which should be considered alongside clinical expertise and judgement.

The diagnostic process leads to a formulation that brings together information from the various assessments and investigations and finally determines the likely cause of the observed clinical changes and sets them in the context of the individual. This is then the basis for making a diagnosis and developing an individualised care plan, based on knowledge of the individual and their surroundings.

6.1 Assessment Process

The section outlines in more detail the key factors to be considered in the assessment process.

1. **History**
    - Nature of the problems, origin, rate of progress, presence of seizures and other associated problems, impact on the person's overall functioning and personality.
    - Particular emphasis on cognitive functions, daily living skills and biological functioning. Comparison with previous assessment results.
    - Presence of psychiatric symptoms such as depression, anxiety.
    - Past history: mental health problems, significant physical and medical history including past and present medical conditions, e.g. diabetes, hypertension, cerebrovascular accident (CVA).
    - Family history: dementia or other mental health problems.
    - Personal history: e.g. developmental history, best level of functioning, life events.
    - Information gathering should be undertaken through a combination of informant interview (preferably with an informant who has known the person well for a period of six months at least) and directly from the person where possible.
2. **Mental state examination:** Observation of level of alertness and any clouding of consciousness, psychomotor activity, mood, thoughts, evidence of any abnormal mental beliefs or experiences, and perceptual abnormalities and memory and other cognitive assessments (see below).
3. **Physical examination:** Where possible a complete physical examination should be undertaken.

    The key issues are:

    - Cardiovascular system – focal deficits, evidence of CVA, etc.
    - Detailed neurological examination (focal deficits, gait abnormalities, speech abnormalities, etc.).
    - Endocrine system: signs of hypothyroidism.

Complete physical examination may be difficult in a significant proportion of individuals. The physical health issues could be addressed in these individuals by a combination of:

- Brief physical checks, e.g. blood pressure, pulse.
- Observations for any evidence of physical health issues.
- Information from the carers.
- The '*OK Health Checks*' (Matthews, 2006)" and Health Action Plans (DH, 2001) can be used to structure this process.

4. **Physical Investigations:**

Recommended routine investigations are:

- Full blood count.
- Urea and electrolytes.
- Blood sugar.
- Thyroid function tests.
- Liver function tests.
- B12 and Folate level.
- Lipid profile.
- Sensory screening – vision and hearing.

Optional tests are:

- Electro Encephalograph (EEG) – if there is evidence of the occurrence of seizures.
- Neuro-imaging: this may not be practical as a routine investigation for the assessment of dementia but may be of value when vascular dementia or other brain lesions are suspected (see Section 6.3 below).

- Electro Cardiograph (ECG) – particularly if anti-dementia medication is to be used or there is evidence of cardiovascular problems.

5. **Environmental assessment** (see also Section 11)

- Quality of the person's physical environment.
- Staffing levels (day and night).
- The mix of people with learning disabilities in the residential and day care settings.
- Quality and quantity of day activities.
- Staff characteristics: attitudes and competence, including consistency of approach.

Key points:

- Carrying out investigations may be difficult in some individuals with Down's syndrome. In this situation, clinicians have to make a decision using information from the history, physical assessment and direct observation on whether investigations are essential or not.
- Mental Capacity Act 2005 and its Code of Practice guidelines must be followed where investigation is considered and the person is unable to consent and cooperate.

6.2 Assessments and information to support the diagnostic process

6.2.1 General points:

- Assessment tools for dementia in the general population are not appropriate for people with learning disabilities. For example, widely-used tools such as the Mini-Mental State Examination (MMSE) (Folstein & Folstein, 2001) assume the pre-morbid level of functioning to have been within the average range.
- Always look for evidence from previous assessments on file that might indicate pre-morbid functioning. Where available, the same test can then be repeated for comparison.
- There is no single battery of assessments universally used by learning disabilities teams in the UK but some useful tools are available.
- There is some guidance available (NICE/SCIE, 2006, Burt & Aylward, 2000).
- The tests used should include direct assessment with the person and questionnaire assessments with well-informed carers. Information should be collected from both carers at home and from carers within the person's day services.
- For some individuals with profound learning disabilities, pre-morbid cognitive ability may be so poor that changes may not be detected by any available standardised testing. Carer reports have to take precedence.
- Whichever battery of assessments is chosen, it should be used longitudinally within the service to enable comparison of performance over time.
- A common problem with the validation with most scales is the small number of people with Down's syndrome and dementia (average between 10 to 12) included in the validation studies for these scales.

Some test batteries contain both direct assessments and informant interviews together. These are dealt with in the relevant separate sections below.

6.2.2 Direct testing with the person with Down's syndrome/learning disabilities

Tests should cover short and long term memory, orientation, communication level, and mood. This is not a complete list. The tools listed below are those most commonly used in services in the UK. Specific recommendations cannot be made until there is more evidence comparing their efficacy.

Neuropsychological assessments

CAMCOG-DS is the neuropsychological assessment part of the CAMDEX-DS (Ball et al., 2006). It is a concise neuropsychological test battery which is based on the CAMCOG, which was designed to meet the need to assess all the cognitive deficits specified in criteria for dementia i.e. memory impairment, aphasia, apraxia, agnosia and disturbance in thinking (executive function). The CAMCOG-DS includes assessments of orientation, language, memory, attention, praxis, abstract thinking and perception, giving individual subscale scores as well as a total score.

Neuropsychological Assessment of Dementia in Adults with Intellectual Disabilities (NAID). (Crayton et al., 1998). The NAID is a battery of very simple tests covering memory, orientation, language and praxis. There is no manual, rather the instructions are in Crayton et al., (1998) and data in Adams & Oliver (2006). This battery takes about 45 minutes to administer. The majority of people with Down's syndrome can attempt most of it. It is said by the authors to be in use in over 30 learning disabilities services in the UK.

Severe Impairment Battery (SIB) (Saxton et al., 1993). The SIB is designed to assess cognitive abilities at the lower end of the range in the general population (age range 51 to 91). There are 40 items and administration is said to take about 20 minutes. It is composed of very simple one-step commands which are presented in conjunction with gestural cues (e.g. 'what's your name?', 'please write your name here', 'what do you call the thing you drink coffee from?' The SIB is divided into scorable subscales, each sampling within the range expected of the severely-impaired individual. The six major subscales are attention, orientation, language, memory, visuospatial ability and construction. There are also brief evaluations of praxis and the person's ability to respond appropriately when his/her name is called (orienting to name). In addition, there is an assessment of social interaction skills. It yields scores out of 100, to assess mild to moderate dementia. There is no cut-off for 'normal' as the test should only be used with people known to be severely impaired.

Test for Severe Impairment (Albert & Cohen, 1992). This is a 24-item test covering eight domains which was designed for people with severe cognitive dysfunction, but not specifically learning disabilities.

Dalton Brief Praxis Test (Sano et al., 1991). This is an abbreviated, 20-item version, of the Dyspraxia Scale for Adults with Down's Syndrome, a 62-item cognitive test of praxis. The BPT tests ability to perform simple, highly practised, voluntary movements in response to a verbal command or imitation. It therefore measures verbal comprehension and motor co-ordination.

Measure of depression

The **Glasgow Depression Scale** (GDS-LD) (Cuthill et al., 2003). A 20-item questionnaire designed for use with people with learning disabilities. It has a three-point Likert-type response scale ('never', 'sometimes', 'a lot'). It has a suggested cut-off score for suspected depressive illness.

Assessments of daily living skills through direct observation of the person

If repeated at intervals, these can shed additional light on changes in skills. There are many to choose from, for example the **AMPS** (Fisher, 2006) (which is limited to Occupational Therapists who have received training in it), or the **ABAS-11** (Harrison & Oakland, 2003).

6.2.3 Informant questionnaires

These should aim to cover those areas of functioning that are known to deteriorate with the development of dementia including: short- and long-term memory, general mental functioning, dyspraxia and dysphasia, daily living skills, personality and behaviour.

CAMDEX-DS informant interview (Ball et al., 2006) is a structured interview specifically designed for the diagnosis of dementia in people with learning disabilities to be carried out, in the absence of the person with learning disabilities, with a relative or carer. The informant should have known the person for at least six months. The interview takes approximately 40 minutes to complete and comprises four parts: person's best level of functioning, cognitive and functional decline, mental health and physical health. The interview asks about current level of functioning and, if a functional limitation is noted, whether this is as a result or not of a slight or great deterioration from previous functioning.

General dementia screening tools

Dementia Questionnaire for People with Learning Disabilities (DLD) – formerly known as the DMR (Evenhuis et al., 2007). The DMR and its recent successor, the DLD, is widely used to assess dementia in adults with learning disabilities in the UK and Europe. It is a screening tool for the early detection of dementia in adults with learning disabilities, completed by carers, consisting of 50 items. There are eight sub-scales: short term memory, long term memory, orientation (making up Sum of Cognitive Scores), speech, practical skills, mood, activity and interest and behavioural disturbance (making up Sum of Social Scores). Evenhuis (1992, 1996) reported that the DMR had sensitivity of up to 100 per cent in identifying dementia and suggested change scores and cut-off scores that might be indicative of dementia. However, Evenhuis (1996) found poor inter-rater reliability for DMR. The main problem with the DMR in clinical practice is to determine the level of learning disability as DMR cut-off scores are based on the level of learning disability. This may be the main reason for Evenhuis to find poor reliability for her scale. Prasher (1997) conducted an independent evaluation of the DMR on 100 adults with Down's syndrome in the UK and, finding poor specificity, suggested modifications to the cut-off scores.

The Dementia Scale for Down's Syndrome (DSDS) (Gedye, 1995). This is designed for use with people with Down's syndrome but may also be useful for people with learning disabilities generally according to NICE (2006). The manual says that its administration is restricted to clinical psychologists and trained psychometricians. It gives a measure of early, middle and late stages of dementia and includes the time course of the deterioration and a differential diagnosis scale. The psychometric property of the DSDS has never been published in a peer-reviewed journal.

Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID) (Deb et al., 2007). The DSQIID is designed to be 'a user-friendly observer-rated dementia screening questionnaire with strong psychometric properties for adults with intellectual disabilities', according to the authors. It comprises 43 questions in three sections. The scoring system overcomes the floor effect in other assessments. It is too early for there to have been independent evaluation of its utility.

Adaptive Behaviour Dementia Questionnaire (ABDQ) (Prasher et al., 2004). This is a 15-item questionnaire derived from the AAMD Adaptive Behavior Scale (Nihira et al., 1974) which is used to detect change in adaptive behaviour. It has been developed to screen specifically for dementia in Alzheimer's disease. It sets out to collect information on how the person compares now to their previous normal level of social functioning. It gives criteria for the presence of Alzheimer's disease and a rating of severity.

Carer burden

Caregiver Activity Survey-Intellectual Disability (CAS-ID) (McCarron et al., 2002) provides a measure of carer burden that can be helpful as a measure of increasing care needs and therefore in determining level of social care required. Carers record the amount of time needed to care for a person with learning disabilities in a 24-hour period, across eight domains such as nursing care, behaviour, supervision and personal hygiene. Care staff require careful instructions to complete it reliably.

Measure of depression

Glasgow Depression Scale Carer Supplement (GDS-CS) (Cuthill et al., 2003). This is a 16-item questionnaire about depressive symptoms, completed by a carer. It is designed to be applicable to people with learning disabilities. No cut-off scores are suggested by the authors, so its usefulness depends on repeat use.

Life events scales

These are useful to gauge whether the presenting symptoms may be attributable to an adverse life event. There are many different ones available or they may be designed by the assessor to cover typical life events occurring in the past two years.

Assessments of daily living skills completed with a carer

If repeated at intervals, these can shed additional light on changes in skills. There are many to choose from, for example:

- **AAMD Adapted Behaviour Scales (ABS), 1974 revision** (Nihira et al., 1974), which is a standardised measure of daily living skills and maladaptive behaviour. It can be useful for assessing those with profound intellectual disabilities, who cannot undertake direct assessments.
- **Hampshire Social Services – Staff Support Levels Assessment** (Hampshire Social Services, 1987) – a 32-item assessment which has excellent behavioural anchors and, therefore, has excellent test-retest validity and inter-rater reliability.
- **Vineland Adaptive Behaviour Scales: Second Edition** (Sparrow et al., 2007) provides analysis of a broad range of skills and behaviour. These include communication, daily living skills, socialisation, motor skills and maladaptive behaviour. Vineland Adaptive Behaviour Scales: The second edition is available in both long and short form – providing either a summary assessment of adaptive behaviours or a more detailed assessment. Semi-structured interview and questionnaire formats make it easier to assess those who have difficulty performing in test situations. It covers 0 to 90 years. It is an American tool and hence some of the items will be unfamiliar to UK users.
- **Adaptive Behaviour Assessment System-II (ABAS-II)** (Harrison & Oakland, 2003) provides a comprehensive norm-referenced assessment of the adaptive skills of individuals aged from birth to 89 years. The clinician can use the ABAS–II to diagnose and classify disabilities and disorders; identify an individual's strengths and limitations; and to document and monitor the individual's performance over time. It provides standard scores that are directly comparable to those found in the Wechsler IQ tests. It too is an American publication.

In looking at changes in adaptive functioning, it is important to assess not only the ability of the person to complete each task, but to probe carefully whether there is a qualitative change in performance of each task.

6.2.4 Who conducts assessments and how are they organised?

A care pathway is needed to guide the multi-disciplinary team through the sequence of events from referral, through screening and diagnosis, to treatment and end of life care. It helps to ensure a co-ordinated approach and effective multi-disciplinary working. Typically, a community nurse conducts the health screen as the first step, in partnership with the GP and/or learning disabilities psychiatrist for necessary blood tests. The assessments of memory, mood and behaviour are often conducted by clinical psychologists but also by other multi-disciplinary team members. AMPS assessments, if used, require special training (usually the preserve of occupational therapists). The role of the learning disabilities psychiatrist is crucial to making the differential diagnosis once all the assessment data have been collected.

Some services have a dedicated or virtual team using an agreed battery of assessments (e.g. McBrien et al., 2005). This may be known as a memory clinic or dementia screening programme. The team should at the minimum include a psychiatrist in learning disabilities, a clinical psychologist and community nurse.

Assessment in rural areas can be made more efficient by use of telemedicine. A telemedicine memory clinic for adults with Down’s syndrome is described by Brown, (2004). Audit of this service showed it to be very helpful in a rural area, acceptable to clients, and saving of travel time.

6.2.5 Where to see people/observations

Careful attention needs to paid to where, when and how to assess individuals. An holistic assessment should include meeting the person being assessed and their carers in their normal living and daytime environments. Cognitive assessments may be more effectively offered in a healthcare or similar setting if this can provide consistency for repeat assessment and provide standardised testing conditions that are free of distractions and give an appropriate context.

6.2.6 Conditions for reassessment

Repeat cognitive assessments need to be rigorously administered and interpreted to take account of normal fluctuations in the assessed person’s performance (e.g. tiredness, mood, good day/ bad day), specific changes (e.g. hearing aids, glasses, medication effects), testing environment changes (place, layout, distractions) and tester effects (skills, experience, relationship with the person with learning disabilities, testing style, administration and scoring anomalies). Ideally the same tester should use the same tests in the same environment using strict criteria for similar administration/prompts, and where possible seeking information from the same informant on each occasion. The standard error of measurement and normal ageing deterioration must also be considered when considering results showing some evidence of cognitive deterioration.

Key points:

- Multi-disciplinary assessment is important.
- Assessments should include direct assessment of the person together with informant based questionnaire/assessments.
- Assessment for other co-morbid conditions is essential.
- Consideration should be given to practical issues in assessment (e.g. location).
- Consider test re-test issues (e.g. different informants/testers).

6.3 Neuro-imaging

Indications

The most consistent structural change in the early stage of Alzheimer's disease is the atrophy of the medial temporal lobe.

People with Down's syndrome have medial temporal lobe atrophy even without dementia. However, normative values have not yet been established, so neuro-imaging currently has limited value in the early diagnosis of Alzheimer's disease in people with Down's syndrome. Its value is mainly to rule out structural lesions other than atrophy (e.g. space occupying lesions). It should, therefore, be used only when the clinical picture suggests the possibility of such lesions.

MRI scans have a number of advantages over CT scans. However the sensitivity of CT scans has improved considerably and the procedure is shorter and less complex than for MRI scans. Deb et al. (1992) report that SPECT scans are more useful than either CT or MRI scans.

Informing and preparing

Neuro-imaging requires explicit informed consent. Detailed information regarding the rationale and the procedure should be given to the patient in an appropriate user-friendly format. Assessment of capacity will be needed to identify whether the individual is able to provide valid consent for the procedure. Where the person does not have capacity best interest principles will need to be applied.

Anxiety about the procedure can be allayed by a visit to the radiology unit and familiarisation with the procedure.

Process

Although with adequate preparation, some people with learning disabilities may be able to go through the procedure without any other interventions, others may benefit from the use of one dose of oral anxiolytic medication such as lorazepam or diazepam an hour beforehand. Some clinicians prefer to use buccal midazolam, which provides rapid and short-term sedation and, therefore, may be given immediately before the procedure. Some individuals may find it difficult to have a scan in spite of all these measures.

Generally, the MRI procedure is longer and more anxiety provoking. In these situations, discussion with the radiologist may be helpful in deciding if CT can be used as an alternative. New generation CT scans are much more user friendly and less anxiety provoking.

If the individual clearly needs neuro-imaging but is unable to co-operate in spite of all these measures, it can be undertaken under general anaesthesia. However, risks and benefits should be evaluated and thorough discussion held with the carers and other professionals to arrive at a best interests decision, if the person lacks the capacity to consent to this specific investigation. If the person has the capacity to consent to the scan it is for him/her to decide whether or not to have the scan.

Sharing the findings

It is important to share the findings of the scan with the person with learning disabilities, carers and other professionals. This should include the nature of the findings as well as implications for the management. This discussion should be clearly documented.

Key points:

- Neuro-imaging (CT/MRI) is not an essential investigation for the diagnosis of dementia in Down's syndrome. It may be of value where other brain lesions/vascular dementia are suspected. New generation CT scanners are as sensitive as MRI scanners and may be more acceptable for people with learning disability.

Section 7 – Decision making/telling people

Disseminating results of assessments

Assessment information may not need to be referred to again for several years, therefore it is important to ensure copies are securely held where they may be found in future. Some services copy their assessment reports to the GP, the residential home or family carer as well as to the person's learning disabilities NHS file. Accessible reports should be considered for the person with learning disabilities.

1. Establishing the diagnosis:

The first step in establishing the diagnosis is to bring the information from different sources together. It is important to combine the assessments of the psychiatrist, psychologist, occupational therapist, community nurse and other relevant professionals with the results of the investigations and physical examination that the GP or psychiatry team may have carried out. The psychiatrist will be in the unique position of reviewing the findings and then arriving at a diagnosis with support from the multidisciplinary team.

2. Multidisciplinary discussion and care coordination:

In the first multidisciplinary meeting, it is important to agree on the care plan at an early stage and identify an appropriate professional to be the care coordinator. This should form part of the Care Pathway.

3. Diagnosis:

For a significant proportion of individuals, it may not be possible to reach a clear diagnosis at an early stage. It may only be possible to have a range of differential diagnoses with a true picture emerging over a period of time. However it is worthwhile remembering that for some individuals at this stage the diagnosis of dementia may be clearly ruled out.
The diagnostic evaluation may need to be repeated regularly, for example, at six monthly intervals.

4. Management:

Irrespective of the degree of certainty attached to the diagnosis, interventions focused on the needs of the individual and management of any risks should be thoroughly discussed in the first care planning meeting and management plans agreed.

5. Sharing the information with the person with learning disabilities:

Steps should be taken to share the information with the person with learning disabilities at all stages and this may involve use of a 'communication passport' and additional input from a Speech and Language Therapist (SALT) to make the information as user friendly as possible. This process should be undertaken as good practice irrespective of the mental capacity of the person concerned. Existing booklets, for example, *The Journey of Life*, *About My Friend* and *About Dementia*, (Dodd et al., 2005 a, b & c), *What is Dementia?* (Kerr & Innes, 2000) and *Breaking Bad News* (Buckman, 1992) are useful resources.

6. Sharing the information with carers:

This will involve giving information on the diagnosis, including likely course and prognosis. Every effort should be made to keep carers informed at all stages. Information should also be shared about the management plans and the rationale behind them. Carers will need support for end of life preparation at a later stage. There are information booklets to assist carers to understand dementia e.g. *Keep Talking about Dementia* (Watchman, 2003) and *Down's Syndrome and Alzheimer's Disease: A guide for Parents and Carers* (Holland, 2004). Useful information can also be found on the Alzheimer's Society website.

Key points:

- Decision-making should be based on multi-disciplinary assessments and discussions.
- When informing carers and people with learning disabilities about the nature of the diagnosis, it is important to bear in mind that the diagnosis is often provisional and a true picture may only emerge after a length of time. This should however not prevent clinicians considering and implementing the necessary steps to deal with the situation as it currently presents.

Section 8 – Additional health/clinical issues associated with dementia

8.1 Epilepsy

Prevalence

Over 80 per cent of people with Down's syndrome and dementia develop seizures (Lai & Williams, 1989). Older people with Down's syndrome (over 45 years) are more likely to have seizures than younger people. A younger age of onset of dementia is associated with a higher risk of developing seizures.

The nature of seizures

The common type of seizures in people with dementia are myoclonic and generalised tonic clonic types though the whole range of both generalised and partial seizures may be seen.

- In the general population, people with Alzheimer's disease generally develop seizures at a late stage of dementia but in people with Down's syndrome seizures can be a presenting symptom. Seizures generally tend to occur earlier in the course of the illness than is found in the general population.
- Epilepsy has two peaks of onset in people with Down's syndrome. The first peak is the development of epilepsy during infancy, the second is associated with the onset of dementia. Therefore, a proportion of people with Down's syndrome who develop dementia will have a previous history of epilepsy, and there may be a change in seizure frequency, pattern or severity with the onset of dementia.
- Frequency and intensity. Myoclonic seizures tend to occur more frequently in the early morning but may progress at a later stage to any time of the day. These are generally mild jerks, although the intensity and frequency can vary considerably. Generalised tonic clonic seizures are more obvious and similar precautions as in people without dementia should be taken.

Investigations and Diagnosis

The diagnosis of epilepsy is a clinical one, and hence the whole range of investigations may not be appropriate for people with dementia, especially complex procedures such as EEG and CT/MRI scans which may cause more distress to the person than is warranted by any benefits. Other investigations such as a full physical examination and blood tests should always be considered. Neuro-radiological investigations and EEGs should only be undertaken if there is a suspicion of intra-cranial growths or other treatable causes of dementia.

Risks

Seizures are associated with the risk of falls, injuries and sudden deaths. The risks increase with poorly controlled seizures, poly-pharmacy and specific seizures such as atonic seizures and nocturnal seizures. It is important to establish a monitoring system so that staff and carers can take responsive action when needed. Staff and carers should be encouraged to maintain regular seizure charts to record the nature, frequency, intensity and duration of seizures, and to complete risk assessments with regard to risk of falls, and injuries.

Management

(a) Drug management – seizures in people with Down's syndrome generally respond to a single anti-epileptic medication. The commonly used medications are sodium valproate and lamotrigine. The advantages of these medications include a broad spectrum of effectiveness and, particularly in the case of lamotrigine, reduced impact on cognitive impairment. Often these medications also act as mood stabilisers. The following good practice principles should be borne in mind.

   i. Promote the use of a single medication whenever possible.

   ii. The treatment goal should be a healthy balance between quality of life and seizure control.

   iii. Unacceptable side effects should not be present and the prescriber should take immediate action if side effects are reported. Careful monitoring of the side effects by staff and carers is the key and it is important for clinicians to explain what to look for in relation to side effects.

   iv. The care plan should cover how to minimize seizure related risks especially in relation to falls and injuries.

(b) Use of rescue medication. The commonly used rescue medications are diazepam rectally or midazolam buccally. The standard doses of both are 10mg. Buccal midazolam at 5mg can be used as a starting dose rather than 10 mg, which minimises the risk for respiratory failure. Midazolam is unlicensed for this use but many people prefer it because administration is easy and more acceptable. Training for staff and carers in the use of rescue medications is essential. The use of rescue medication should be documented in the person's care plan.

(c) Side effects. The main side effects of sodium valproate are weight gain, gastro-intestinal problems, and a negative impact on cognitive function. The main side effects of lamotrigine are skin rashes and gastro-intestinal side effects.

(d) Do's and dont's.

   - Do not use rescue medications unless they are clearly indicated and documented in the care plan.
   - Do train carers and provide appropriate information.
   - Remember that quality of life has a higher priority than total seizure freedom.
   - Remember that the presence of active epilepsy should not limit community participation.
   - Do monitor seizures and the side effects of anti-epileptic drugs.

Key points:

- The occurrence of seizures for the first time is very common in people with Down's syndrome who have developed dementia.
- The use of a single anti-epileptic medication should be encouraged.
- Careful monitoring is required of seizure patterns and side effects of medication.

8.2 Pain

Poor recognition and treatment of pain in people with learning disabilities and dementia is common (Kerr et al., 2006). Many issues contribute to the low level of pain recognition. These include:

- staff attitudes towards, and experience of, 'behaviour that challenges'.
- diagnostic over-shadowing.
- problems with communication.
- beliefs about pain thresholds.
- the impact of past treatment on willingness to complain of pain.
- the use of temporary agency/bank staff who are less likely to observe a change in a person's level of distress.

Sometimes staff are not sufficiently aware that people with learning disabilities who are getting older will experience painful conditions, such as arthritis, that can accompany older age. Difficulties are sometimes ascribed to the dementia rather than being a consideration of whether people are in pain.

Research and practice both indicate that there is inadequate training of staff at all levels in all professions about dementia and learning disabilities and, in particular, about the pain management needs of people in this group. There is also little use of pain assessment and recognition tools. There is a range of effective pain/distress tools available for staff and carers to use to identify pain or distress in people with learning disabilities and dementia. Tools to consider include the Abbey Pain Scale (Abbey et al., 2004) and the DisDAT (Regnard et al., 2007).

Key points:

- Pain recognition and management for people with learning disabilities and dementia is often very poor.
- Diagnostic overshadowing is a frequent occurrence, and staff are often unaware of the range of painful health conditions that may present with increasing age.
- There are tools available to help staff and carers identify pain in people with learning disabilities.

8.3 Sleep disorders

People with learning disabilities have a high prevalence of sleep difficulties which worsen with age and with associated conditions such as dementia (Espie, 2000). The sleep disturbances in dementia typically include a reversal of the sleep-wake cycle (sleeping during day time and wandering around at night time) and a reduction in the slow wave sleep which may be due to the loss of cholinergic neurons.

Clinicians should exclude the following treatable conditions or situations before considering biological attributes (loss of cholinergic neurons):

- Co-morbid mental health problems including depressive illness.
- Psychological issues, e.g. fears, anxiety.
- Alcohol/substance misuse.
- Physical health problems, e.g. pain, epilepsy, heart-failure, respiratory disorders, nocturnal enuresis.
- Side-effects of medication, e.g. stimulant drugs.
- Poor sleep hygiene – uncomfortable bed, noisy household, poor light and temperature adjustment, late evening coffee and recent change in the environment.

People with learning disabilities and dementia should have a routine assessment of sleep hygiene as in the general population. Associated factors can be easily overlooked, especially in people with learning disability who may have poor communication skills. The assessment and the sleep history from the carer should include the following:

- The person's sleep pattern.
- Information on sleep hygiene, bedroom and bedtime routine.
- Onset, duration and nature of the problem (e.g. difficulty in going to sleep, frequent awakening, early morning insomnia, motor activities, snoring during sleep and evidence of daytime sleepiness).
- Effect of insomnia on the person and others including family and/or carers.
- Past sleep difficulties and previous treatments.
- Psychiatric and medical diagnoses (including epilepsy).
- Current medications.
- Family history of sleep problems.
- Risks associated with sleep difficulties, e.g. wandering behaviours or falls.

Management

- Treat any associated physical and/or psychological problems.
- Use non-pharmacological strategies (sleep hygiene).
    - Encourage daily activities and if possible exercise.
    - Avoid day time napping.
    - Reduce caffeine and alcohol intake before bed time.

  - Eliminate factors that impede sleep (watching TV into the early hours).
  - Use the bed just for sleeping.
  - Set and maintain a regular routine of rising and retiring at the same time everyday.
  - Ensure sleeping environment is conducive to sleep (comfortable bed, quiet, dark and at the right temperature).

If the above approaches do not produce any significant benefits and the risks continue, a pharmacological approach may be considered along with non-pharmacological approaches.

The pharmacological approaches include the use of:

- Benzodiazepines (Temazepam).
- Z drugs (Zopiclone and Zolpidem).
- Melatonin (Sajith & Clarke, 2007) – recently licensed under the name of Circadin in patients over the age of 55 (there is an age-related decrease in endogenous melatonin secretion which contributes towards age related insomnia).

The choice of medication should depend on the individual patient's needs and should be based on NICE Guidelines (2004) recommendations:

- Use benzodiazepines as the first line approach, but be careful about the long-term effects of such drugs.
- Consider use of z drugs as a second line alternative.
- There are no differences in the efficacy of z-drugs and if one of them is not effective the others should not be used.
- Switch from one z-drug to another only if there is an adverse effect directly related to that particular drug.
- Use the minimum effective dose.
- Use medication on an 'as and when necessary' basis (every second or third night if required) rather than on a regular basis.
- Do not continue hypnotics for longer than four weeks.
- Discuss the discontinuation with the person and the carer, and taper and stop it very gradually.
- Warn the person and carer of withdrawal symptoms and rebound insomnia.
- Advise on adverse interactions with alcohol and other sedative agents.
- Do not use in hepatic failure, chronic respiratory diseases and people who have a history of substance misuse.
- Be mindful of the side effects including day-time sedation, falls and sundowning (confusional state in the evening).

Key points:

- Sleep difficulties are commonly experienced in people with learning disabilities as they get older and/or develop dementia.
- Assessment should include ruling out co-morbid mental health problems, substance misuse, physical health problems and poor sleep hygiene
- The management of sleep difficulties in dementia should be based on non-pharmacological approaches including good practice of sleep hygiene.
- Medication should only be used if other approaches have failed or risks are significant.
- NICE recommendations should be adhered to if pharmacological approaches are used.

Section 9 – Conceptual understanding of the dementia process

Conceptual understanding of the psychological and social consequences of dementia is essential for senior staff to guide timely decision-making about interventions and approaches to individuals/service developments. The social model (NICE/SCIE, 2006) proposes that whilst people with dementia have an impairment, they may be disabled by the way they are treated by or excluded from society. The advantages of this framework are that carers and staff will understand that dementia is:

- not the fault of the individual;
- the focus is on the remaining skills rather than losses;
- the individual can be fully understood (their history, likes/dislikes, etc.);
- the influence of an enabling or supportive environment;
- the key value of appropriate communication;
- opportunities for stress-free and failure-free activities.

This means that the responsibility to continue to reach out to people with dementia lies with people who do not have dementia. Carers and staff need to change their approach to 'go with' the person and their continuing changes. Brawley (1997) concluded that 90 per cent of the catastrophic behaviours in dementia are induced by carers or the environment.

Downs et al., (2006) show how different models e.g. as a neurological condition; as a neuro-psychiatric condition; as a normal part of ageing; and seeing dementia from a person-centred perspective, can be used to understand dementia. One approach to understanding dementia and its psychological consequences has been put forward by Buijssen (2005). He proposes two laws of dementia, and asserts that by understanding them, and their consequences, we have a framework to understand and respond to people appropriately.

Law 1: Law of disturbed encoding

In this law, the person is no longer able to transfer information successfully from their short-term memory and store it in their long-term memory. This means that the person is unlikely to remember things that have just happened to them.

The main consequence of disturbed encoding is that the person is unable to form any new memories for the things they experience or for things they are told. This means that they experience:

- Disorientation in an unfamiliar environment.
- Disorientation in time.
- The person asks the same questions repeatedly.
- The person quickly loses track of conversations.
- The person is less able to learn anything new.
- The person easily loses things.

- The person is unable to recall people whom they have recently met.
- Appointments are quickly forgotten.
- The person experiences anxiety and stress.

Law 2: Law of roll-back memory

Long-term memory contains all the memories that have been acquired starting with the most recent memories, and working back toward childhood memories. When dementia develops, the person will be less able to form any new memories after this time. At first their long-term memories will remain intact, but as dementia progresses, long-term memories will also begin to deteriorate and eventually disappear altogether. Deterioration of memory will begin with the most recent memories and will progress until only memories of early childhood remain, hence memory can be said to be ‘rolling back’.

The consequences of roll-back memory are:

- Loss of daily skills such as using kitchen appliances.
- Memory loss for events, beginning with the most recent, e.g. last holiday.
- Decreased social skills and increased inappropriate behaviour.
- Decreased vocabulary and inability to find words.
- Disorientation towards people, e.g. inability to recognise family and relatives.
- The person may begin to have ‘flashbacks’ and see people from their past.
- Self care skills will begin to deteriorate.
- Changes in personality.
- The person believes that they are younger and that time has actually ‘rolled back’.

Key Points:

- The social model gives a conceptual model for staff and carers to understand dementia.
- Understanding what happens as dementia progresses and its consequences gives a framework to understand and respond appropriately.

Section 10 – Philosophy of care

10.1 Individualised philosophy of care

Excellence in dementia care requires staff and family carers to understand and know the person, understand dementia and its consequences for the person, and consequently to be able to think ahead and predict ‘stressors’. They need to adapt their approach to ensure that the person with dementia has a stress-free, failure-free but individualised care that is consistent but without time pressures. This needs to be incorporated into the person’s Person Centred Plan and Care Plan.

This can only be achieved by having a thorough knowledge and understanding of the person and their history. This can be aided by the use of Life Story work (Gibson, 1994).

Staff and family carers need to be clear about what they are trying to achieve. This is not the time for learning new skills, achieving goals or facing change. They need to consider the person’s happiness, comfort and security. The focus of care should move away from targets to quality of life. Staff and carers need to remember that people compensate for their deterioration in functioning by making greater use of remaining abilities (e.g. earlier memories). This may mean that the person finds comfort in activities and objects from their childhood. Roll-back memory may also mean that the person is not oriented to the present day. Care needs to be taken not to challenge the person’s beliefs repeatedly as this will add to their stress.

Successful support includes not only the social aspects of the person’s life, but an increasing awareness of the physical consequences of getting older and additionally having dementia. Care needs to be taken to ensure that diagnostic overshadowing does not occur, where all changes are attributed to the dementia. Research (e.g. Kerr et al., 2006) indicates that pain recognition and management is extremely inadequate in this group of people. All people should have comprehensive health checks and a Health Action Plan, updated annually at minimum.

10.2 Family carers

Family carers need specific emotional and practical support. Many family carers find the diagnosis of dementia traumatic, as it may bring back emotions surrounding the birth, life expectancy and disability of the person. Where the family are the main carers, they must be offered a comprehensive Carer’s Assessment. Many carers, particularly the parents of people with Down’s syndrome, may themselves be at risk of developing dementia or other age-related conditions.

Services need to be sensitive to the needs and beliefs of carers, and to see things from their perspective. Some carers believe that it is their duty to care and may find it very difficult to accept support and help into their own home. Carers need to have prompt access to appropriate information about supports and resources available, including short breaks (both within and away from the home), individualised budgets and direct payments, and aids and adaptations.

Appropriate use of the National Framework for Continuing Health Care (DH, 2007) should be made to ensure that funding of health and social care is fair and transparent.

Staff need to be very sensitive to the small number of carers who cannot cope with seeing their family member deteriorating, and may opt out of being involved. Life Story work is one positive way of engaging family carers in the care.

Carers often need a great deal of support to prepare for the eventual death of the person they are supporting.

10.3 Paid care staff characteristics

Staffing numbers need to be appropriate to ensure the person is safe and that staff are able to meet the person's changing needs. In practice, this means that staffing levels will need to increase as the dementia progresses. By mid-stage dementia, people usually require waking night staff to ensure safety, and often an increase in staffing levels to manage self care and to respond to the distress that people with dementia often exhibit.

Excellent dementia care can only be provided by a consistent staff group; agency staff, unless very well known to the person, should always be avoided as this can add to the person's stress level. Staff will need to have access to regular training and resources to meet the varied demands of caring for people with learning disabilities and dementia. There is a range of resources available including the *Down's Syndrome and Dementia resource pack* (Dodd et al., 2002), the *Down's Syndrome and Dementia Workbook for Staff* (Dodd et al., 2006) and a range of DVDs currently being made by the Down's Syndrome Association. In our experience, the best care is provided by staff who are flexible in their approach, deal with changing situations with compassion and humour, cope with the person's declining abilities and can reach out to the person.

Staff need support to cope with the deterioration in the person with learning disabilities and dementia and to prepare for the eventual death of the person they are supporting.

10.4 Working with other residents

The effect on other people with learning disabilities of seeing a peer deteriorate is rarely considered. Helping peers to understand dementia can reduce their distress and help them to be more considerate and understanding of the changes occurring with the person with dementia, in some cases helping to prevent placements breaking down.

Resources now exist to help explain dementia to adults with learning disabilities. Dodd et al , (2005 a, b & c) published three booklets (1) *About Dementia,* (2) *About My Friend* and (3) *The Journey of Life* to support this aspect of work. These booklets explain dementia in terms of the lifecycle. They can be used individually or in a group setting, as long as this is supported by confident staff/carers. Both Lyngaard & Alexander (2004) and Dodd (2008) have reported on successful short courses for people with learning disabilities living with someone with dementia.

10.5 Ethnicity issues

Ethnic background has been shown to affect the nature of care giving practices, carer stress and help seeking in people with dementia and their carers (Iliffe & Manthorpe, 2004). In people with learning disabilities in general there are significant differences in the nature of care giving, carer stress and carers' psychological and physical burden (McGrother et al, 2002, Devapriam et al.,2008). Contrary to the popular view, very few enjoy the support of extended family networks (Hatton et al., 2002). Material disadvantage, lack of informal networks and high need for services are some of the key issues related to people with learning disabilities from an ethnic minority background (Hatton et al., 1998).

However it is not clear how these processes are affected when an individual with learning disability from an ethnic minority background develops dementia. Clinical experience suggests that the lack of availability of culturally appropriate respite and home care provision cause difficulty. Lack of awareness of services and language barriers may prevent people from ethnic minority communities from receiving appropriate levels of support. This can result in significant distress for both the person with learning disabilities and their carers.

This highlights the need for more cohesive action by health, social care and voluntary sector services together to improve access to and the experience of services by people with learning disabilities and dementia from Black and Minority Ethnic (BME) communities. The plan of action should include the following:

- A clear strategy for staff training on diversity.
- Proactive measures to develop partnerships with community and voluntary organisations.
- Facilitation of active participation by people with learning disabilities and carers from BME background in the stakeholder discussions about the development of culturally appropriate services for people with learning disabilities and dementia.
- Direct payments and individualised budgets may provide an opportunity for increasing the range of culturally appropriate services available.
- Further research is required to understand the views of people with learning disabilities with dementia from BME communities and their carers about culturally appropriate way of meeting their needs.

Key Points:

- Staff and carers need to ensure that people with learning disabilities and dementia have stress-free, failure free and consistent care.
- This approach leads to a more supportive environment and lower levels of behaviour and distress.
- Services need to review regularly the support given to people with learning disabilities and dementia, particularly the amount of staff support provided both during the day and through the night.
- Capable dementia services need to be aware of, and meet the needs of, the person with dementia, those of staff and carers and other people with learning disabilities who live with the person, including those from BME communities.

Section 11 – Environments

This section of the guidance focuses on how carers and services can create capable environments that enhance the quality of life for people with learning disabilities and dementia.

11.1 Where should people with learning disabilities and dementia live?

Janicki and Dalton (1998) proposed a potential pathway that someone might follow after a diagnosis of dementia. This we have adapted to reflect the current position of the Care Services Improvement Partnership and the Department of Health, with the options given in order of preference for people with learning disabilities and dementia.

Preferred option:

'Dying in place' where the person can stay where they are currently living with appropriate supports adapted and provided. This means that the person stays with what is familiar in their long-term memory.

Compromise option:

'Moving to more specialist learning disability provision' where the person has had to move from their current home, but moves into provision supported by learning disabilities services.

Least preferred option:

'Referral out of learning disability services' where the person will be moved to services for older people, either residential or nursing.

By staying where they are, the person will stay with familiar people (family, peers, familiar carers) and in an environment that they know. Although their needs will change as the dementia progresses, every effort should be made to maintain their home life. This may necessitate environmental changes and adaptations to support the person, increases in staffing levels and careful thinking about the supports required (see Sections 11.2, 12, 14 and 16).

In some instances it is not possible to maintain the person in their existing home:

- It may be that the design of the building is inappropriate and cannot be changed.
- The person may be being cared for by older family carers who may not be able to continue caring as the needs of the person increase, or their own health changes.
- Risk assessments should be completed regularly to ensure that the environment remains safe for both the person and their carers.
- Staff may reach a 'tipping point', where there are insurmountable problems with the placement (often the quality of life for the other residents or staff issues).
- Funding may be an issue.

If the person has to move then the most appropriate provision may be a specialised learning disability service which is able to meet the person's increasing health needs and provide palliative care (Thompson & Wright, 2001). However, although some moves may be of benefit to the person, the majority of the evidence suggests that moving increases the rate of decline and can precipitate deterioration in health and behaviour and lead to an earlier than expected death (Wilkinson et al., 2004). Multiple moves must always be avoided. The use of an advocate or an Independent Mental Capacity Advocate (if the person has no family) may assist in decision-making, particularly where the person is lacking capacity (Mental Capacity Act, 2005).

If the person has to move, it is vital that other opportunities for continuity, e.g. day activities, leisure and social opportunities which are in the person's long-term memory, are maintained. Maintaining social networks will also involve working with the person's peers to help them understand dementia and the changes in the person. There is a range of materials to assist with this (Dodd et al., 2005 a, b & c). See Section 10.4.

11.2 What are the features of a capable environment?

There is a wealth of literature within general dementia care, and a smaller amount within the learning disabilities arena about the importance of the environment in enabling the person with dementia (Kerr, 1997, 2007; Dodd et al., 2002, 2006). Most environments where people with learning disabilities live are not 'dementia-enabled'. Adaptations will need to be made to ensure that the environment does not add more stress. Many of the adaptations required are not too expensive but can have a very positive effect on the quality of life for the person with dementia.

Environments need to be:

- Calm.
- Predictable and make sense.
- Familiar.
- Suitably stimulating.
- Safe and risk assessed.

Problems are created by the use of varied floor colours and textures, having steps and stairs, having patterned carpets, mirrors, installing lights that causes shadows, or installing modern fixtures and fittings that are not within the person's long-term memory. Many of the problems can be solved by the use of appropriate colour, e.g. red toilet seats, and painting toilet doors red, in order to aid orientation.

As the dementia progresses, staff need to be aware of safety issues, especially in relation to access to the kitchen, roads, hot water and electrical appliances. Creative solutions can cost very little, but make a significant difference to the person, e.g. purchasing an old-fashioned push-button TV, rather than using a remote control. Care needs to be taken when refurbishing a home for a person with dementia to ensure that it is equipped with furnishings and equipment that are familiar to the person and their rolled back memory.

Key Points:

- Wherever possible, people with learning disabilities and dementia should remain in familiar environments.
- If the person has to move, then this should be within a learning disabilities service.
- Environments can be adapted to make them dementia friendly and enable the person with dementia to make sense of where they are living.

Section 12 – Meeting changing needs/interventions

12.1 Overview of approaches to intervention

It is important that the diagnostic process, which is often quite lengthy, does not preclude interventions being implemented that can alleviate difficulties being experienced by the person with possible dementia, or minimise the impact of changes being felt by the person's carers and/or peers. Regardless of the eventual outcome of the diagnostic process, help and support are needed to address current areas of need, and this will have to be tailored to individual circumstances, particularly the person's existing health, disability and social circumstances.

Most of the interventions likely to be required are ones that are not specific to dementia, but are core to the work of learning disability professionals i.e. provision of accurate and timely information, interventions to maximise and maintain independence, skills and health, and at the same time to promote safety, comfort, quality of life and dignity. Reducing excessive demands and simplifying routines are key, while minimising avoidable changes, especially any exclusion from appropriate services. It is worth noting, however, that where someone is in a totally unsuitable environment for their needs (e.g. isolated or bullied) and may be depressed, then a change can be very beneficial and aid the diagnostic process.

The use of Person Centred Planning (Valuing People, 2001) and the Care Programme Approach (DH, 2008) provide the same overall framework for individualised planning and care coordination as for anyone else with a learning disability and changing or complex needs. All care planning, of course, has to be in the context of the individual and their family's religious/cultural context. Consent, capacity and risk management issues need to be considered at all stages, alongside the need for advocacy, advance directives, living wills and other mechanisms to facilitate best interest or complex decision making, according to changing legislative frameworks and ethical practice.

12.2 Meeting changing needs

As the dementia progresses, the emphasis of care changes from enabling the person to maintain their skills with support to increasingly taking on tasks for the person with dignity and respect.

At the early stage of dementia, this will involve reminding the person of the day, time, place; simplifying routines and reducing choices; introducing memory aids such as diaries, timetables and objects of reference; simplifying communication, and using additional cues and prompts.

As the dementia progresses, care changes to trying to preserve abilities for as long as possible using techniques of reminiscence, identifying favourite activities and strengths, finding failure-free activities. At this stage, health monitoring becomes essential. Attention to weight, adequate nutrition and hydration, physical health including epilepsy continence, pain and mobility are all vital.

Towards the end of the person's life, swallowing and dysphagia, skin and pressure sore care, moving and handling all become paramount. Even at the end stage of dementia it is important that the person has positive interactions throughout their day (Sharp, 2007). The tasks of daily living often become the activities of the day, and should be pleasurable and enjoyable for the person. Appropriate touch and verbal interaction are essential.

12.3 Understanding behaviour in people with learning disabilities and dementia

By implementing the philosophy of care outlined earlier, many of the difficulties that can occur in dementia can be prevented or minimised. However, there will often be times when people present with behaviours that staff or unpaid carers find difficult to understand and respond to. In all cases the same high standards of assessment of the behaviour and subsequent intervention should occur as outlined in *Challenging behaviour: A unified approach* (RCPsych, BPS, RCS & LT, 2007). This includes ensuring that any intervention addresses the person, the environment and the interaction between the two. As the report says:

> *'A comprehensive assessment should address: a functional assessment of behaviour, underlying medical and organic factors, psychological/psychiatric factors. Detailed functional assessment and diagnosis are both integral features of an assessment of challenging behaviour and should lead to a clear formulation of the presenting problem.*
>
> *Interventions should be delivered in a person-centred context and a framework of positive behavioural support. They should include proactive and reactive strategies. Interventions described include psychotherapy, communication, positive programming, physical and/or medical and psychopharmacological'.* (p.10)

Additional aspects, related to the dementia, may need additional consideration in the assessment of the behaviour of concern:

- The problem behaviour may be transitory to the current stage of the person's dementia and not need an intervention. Similarly, it may be possible to withdraw interventions if/when the person's skills and behaviour change again.
- The situation should be viewed through the eyes of the person with dementia, i.e. their current reality. Continual correction by staff/carers of a false reality (e.g. person with dementia asking when a dead parent will visit) will not reduce their immediate confusion and distress.
- The behaviour should be viewed as an attempt by the person to communicate or to make sense of a bewildering environment (e.g. GP practice waiting room is confused with an airport lounge).
- The behaviour may be an exacerbation or return of previous behaviours. If the person had difficult behaviours/personality traits previously, these may return/re-occur with roll back memory.
- The behaviour may be caused by a return to a long term memory that is now inappropriate, e.g. childhood urinating outdoors in the countryside whilst on long walks.

- There may be an underlying neurological change, e.g. taste bud changes leading to food fads and a liking for stronger flavours, refusing baths resulting from problems with depth perception and/or stepping into the bath.
- Simple and practical solutions may work, e.g. a net to catch items thrown out of the window.
- Simple environmental alterations may alter the behaviour e.g. removing a mirror.

There is a good description of many of the most common problem behaviours seen in dementia in Dodd et al. (2002) *Down's syndrome and dementia resource pack.*

Throughout the progress of the dementia, any changes should be clearly documented. There are tools available to facilitate this, e.g. *Down's syndrome and dementia workbook* (Dodd & Kerr, 2006), and care mapping approaches (Brooker & Surr, 2005).

12.4 Specific interventions

Specific techniques and interventions usually available within older adult services are useful, e.g. reminiscence work, expertise in ageing health issues such as arthritis and pain management, reducing falls, aids and adaptations. Conceptual understanding of the underlying dementia process, drawn from older adult research, is usefully applied to adults with learning disabilities and dementia. One such approach is described below and followed by some other dementia specific approaches beginning to be applied within learning disabilities services. Non-specific interventions that have also proved beneficial are then overviewed.

Very few dementia specific interventions are commonly available within learning disability settings. One such intervention which is being introduced more widely is Dementia Care Mapping (Brooker & Surr, 2005). This observational tool looks at the care of people with dementia and uses approaches of room management and momentary time sampling that were commonly used in learning disabilities services in the 1980s. The 'map' can assist with the development of person-centred care. It usually involves one or two trained 'mappers' sitting in areas such as a lounge or dining area and observing what happens to people with dementia over the course of a typical day. Results are analysed and fed back to the care team, and, along with a report summary, aids person-centred care. This is a copyrighted tool and can only be delivered by staff trained at Bradford University (Basic Mapper three-day course 'Learning to Use Dementia Care Mapping' and Advanced and Trainer courses are available).

There are many useful non-dementia-specific interventions available. All depend on competent staff assessing and intervening as appropriate at whatever level is required (i.e. individual/family/service). The main interventions are outlined in Figure 2: Summary of useful interventions to improve the care of people with dementia.

A summary of the people/services that might be involved/required in a comprehensive service and might participate in the interventions described are listed in Figure 2:

Figure 2: Summary of useful interventions to improve the care of people with dementia.

Physical Health	■ Health checks leading to a Health Action Plan and any treatment. Tools available e.g. OK Health Check (Matthews, 2006) End of Life support - Gold Standard Framework (Thomas & DH, 2005). ■ Management of weight. ■ Management of pain.
Mobility	■ Strategies to maintain mobility, promote exercise, address posture especially regarding respiratory function, correct gait and reduce the risk of falls. Consider equipment needed and promote safe manual handling. ■ Control of pain and discomfort. ■ Pressure area care. ■ Treatment of any difficulties of motor function, adaptation and teaching of skills to include compensatory techniques.
Eating/Drinking	■ Eating and drinking, strategies for maintenance of adequate oral intake in a safe manner. Swallowing assessments, eating programme with dietetic advice and advice regarding posture. ■ Diet to reduce risk of constipation. ■ Dysphagia management.
Continence	■ Aids/adaptations. ■ Help to maintain continence.
Communication	■ Strategies to improve communication. ■ Use of a 'communication passport' i.e. information on how best the person receives information and expresses themselves. ■ Use of objects of reference, pictures. ■ Environmental signposting/signs and symbols
Self Help Skills	■ Modeling/guiding/prompting/pacing. ■ Adding supports to maintain self-care and domestic skills. ■ Adaptation of the environment to aid daily living.
Occupation/Activity	■ Sensory stimulation (including Snoezelen). ■ Aromatherapy. ■ Music and art therapy. ■ Support for people to engage in activities. ■ Other failure free activities e.g. looking at magazines, carers sitting with the person describing what is happening.
Orientation/ Confusion	■ Reminiscence. ■ Life story work.
Mental Health and Behaviour	■ Anxiety Management. ■ Relaxation techniques. ■ Promotion of positive behaviour and feelings of self esteem. ■ Observations to gain an understanding of behaviours. ■ Functional analysis and setting up of programmes for challenging behaviours. ■ Setting up of crisis intervention plans.

Physical Health

GP.
Community Learning Disability Nurse.
Practice Nurse.
Speech and Language Therapist.
Physiotherapist.
District Nurse.
Occupational Therapist (Learning Disabilities).

Level of Functioning

Clinical Psychologist.
Occupational Therapist (including Learning Disability and Social Care Services).
Physiotherapist.
Speech and Language Therapist.

Mental Health and Behaviour Challenges

Community Learning Disability Nurses.
Psychiatrist.
Clinical Psychologist.
Occupational Therapist (Learning Disabilities)

Support for Carers, Family and Peers

Community Nurse Learning Disability.
Social Worker/Care Manager.
Clinical Psychologist.
Psychiatrist.

Future Planning

Social Worker.
Voluntary agencies.

– Mencap.

– Down's Syndrome Association.

– Carers groups.

– Alzheimer's Society.

– Advocacy agencies.

– Religious groups.

Key Points:

- It is important to utilise the skills of the multi-disciplinary team to support the individual with learning disabilities and dementia.
- Other agencies as well as those in the statutory sector can help to support the person and their carers.

Section 13 – Medications

13.1 Anti-dementia medications in people with learning disabilities and dementia

The degeneration of nerve cells in the brain due to the degenerative process leads to a reduction in neurotransmitters (a group of chemicals which have an important role in the transmission of signals between nerve cells). The neurotransmitter acetylcholine is particularly affected in Alzheimer's disease. Acetylcholine plays a key role in the way the brain processes and consolidates information.

All the anti-dementia medications, with the exception of memantine, increase the level of acetylcholine available for transmission of nerve signals by delaying its breakdown. This is achieved by inhibiting an enzyme – acetylcholinesterase – which is responsible for acetylcholine breakdown, a function that normally the human body relies on to maintain the balance of chemicals in the brain. Another enzyme involved in the breakdown is butyl cholinesterase and one of the anti-dementia drugs (rivastigmine) inhibits this enzyme in addition to its effect on acetyl cholinesterase.

- Acetylcholinesterase inhibitors are used mainly in Alzheimer's type dementia.
- NICE guidance (2007) recommends their use in the middle stage of dementia to delay the progress of the illness. However, NICE has restated that the difficulty in staging dementia in people with learning disabilities should not disadvantage them, thus introducing some flexibility for prescribers.
- These medications can also be used in the management of behaviour/psychological problems in people with Alzheimer's dementia and Lewy body dementia where psychological/environmental measures alone are not successful.
- Although there is no conclusive evidence that these drugs are effective in people with learning disabilities, the available evidence suggests that they may improve the quality of life for the person and their carers.
    - A study by Prasher et al. (2005) showed that people who were treated with rivastigmine had less decline over 24 weeks in global functioning and adaptive behaviours.
    - A placebo controlled trial of donepezil (Prasher et al., 2002) showed that the improvement at 24 weeks was statistically non-significant. The sample size of the study was too small to explore the efficacy in the subgroups of mild to moderate disease.
    - Lott et al. (2002) in their open label study on donepezil found that treatment resulted in significant improvement in scores on the Down Syndrome Dementia Scale (Gedye, 1985). However, there were methodological drawbacks.
    - Prasher et al. (2003), in their open label study on donepezil treatment for people with Down's syndrome, found that treatment with the anti-dementia drug was associated with initial improvement in global functioning and adaptive

behaviours. Follow up at 104 weeks found that, whilst there was deterioration in both treatment and control groups, it was significantly less in the treatment group.

- At present there is no known cure for Alzheimer's dementia. Use of anti-dementia drugs at best may improve global functioning or reduce the rate of decline significantly. The natural progression of the disease may be delayed for a period by the medication but ultimately will continue. Eventually tolerance to the medication may decrease with an increased susceptibility to side effects. At this point medication will need to be discontinued. It is therefore imperative to make it clear to carers and service users at the outset that the medication will be withdrawn at some point in the future.
- The effect/side effects of the medication should be monitored closely.

13.2 Psychotropic and other medications in people with learning disabilities and dementia.

Use of psychotropic medications to control behaviour problems in dementia should only be considered as a last resort. The vast majority of people with learning disabilities and dementia with behaviour problems can be managed with environmental and other psychosocial approaches which are detailed in this guidance. Every effort should be made to carry out a thorough assessment to determine the reasons for such behaviours and take the necessary remedial actions (BPS, 2004; RCPsych, BPS, RCS & LT, 2007).

In spite of the use of environmental and psychological interventions, there may be a small group of individuals with dementia who may benefit from use of one of the psychotropic medications. The following are examples where a use of a low dose of medication may be considered:

- The presence of delusions and hallucinations causing significant distress may warrant the possible use of antipsychotic medications.
- People with severe depressive symptoms may benefit from an antidepressant treatment.
- In addition, there will be a small number of people with behaviour problems where clinicians would consider psychotropic medications to minimize the risk to the individuals themselves or other people. In these situations medications are considered as an option due to one of the following reasons:
    - *The behaviour is continuing with significant risk to self or other people in spite of the use of the psychosocial/environmental approaches;*

or

- *where the risk is imminent and it is not practically possible to achieve the risk reduction with any other measures immediately.*

The NICE guideline on dementia (NICE, 2006) states that 'people with Alzheimer's disease, vascular dementia, mixed dementias or Dementia with Lewy bodies with severe non-cognitive symptoms (psychosis and/or agitated behaviour causing significant distress) may be offered treatment with an anti-psychotic drug after the following conditions have been met'. The conditions listed are:

Figure 3: Example of a pathway for the use of acetylcholinesterase in the management of dementia in people with learning disabilities (Bhaumik & Gangadharan, 2008).

Indications:

1. Alzheimer's type of dementia (with or without behaviour problems)
2. Lewy body dementia presenting with behaviour problems

Follow the AAMR guidelines:

- Consider the risks and benefits of treatment with anti-dementia drugs.
- Discuss the risks and benefits with users/carers.
- Gain user/carer consent for a trial of treatment and ensure compliance with medication.
- Carry out electrocardiogram (ECG) if necessary.

Identify key problem areas using one or more of the following scales:

- Dementia Questionnaire for People with Learning Disabilities (DLD).
- Vineland Adaptive Behaviour Scale (Vineland).
- Adaptive Behaviour Scale (ABS) Part 1.

Begin treatment with donepezil, galantamine, or rivastigmine at a minimum possible dose. **Monitor closely** for any adverse drug reactions – establish telephone link with carers.

Reassess the patient after 4 weeks:

- Monitor serious adverse drug reactions; if present stop drug.
- Consider increasing the dose if needed.

Continue to monitor closely for:

- Clinical improvement.
- Any adverse drug reactions.

Reassess clinically at end of 12 weeks and 24 weeks:

- Re-assess key problem areas using DLD, ABS Part 1, Vineland.
- Stop drug treatment in those who show no benefit at 24 weeks despite the use of maximum tolerable dose.

Continue treatment in those who show benefit and reassess at the end of 48 weeks: Re-assess key problem areas using DLD, ABS Part 1, Vineland.

If treatment is continued beyond 48 weeks:

- Continue to monitor patient at 6-month intervals using the rating scales
- Advise carers about reasons for possible discontinuation of treatment in the future, e.g. progression to late stage of the disease.

1. There should be a full discussion with the person with dementia and/or carers about the possible benefits and risks of treatment. In particular, cerebro-vascular risk factors should be assessed and the possible increased risk of stroke/transient ischaemic attack and possible adverse effects on cognition discussed.
2. Changes in cognition should be assessed and recorded at regular intervals. Alternative medication should be considered if necessary.
3. Target symptoms should be identified, quantified and documented.
4. Changes in target symptoms should be assessed and recorded at regular intervals.

It is important to follow good practice principles (Deb et al., 2006; Tyrer et al., 2008) in treating people with learning disabilities and dementia:

- For people with learning disabilities and dementia, any such treatments should be considered on a best interest basis if the person is unable to provide informed consent.
- People with learning disabilities and dementia may be unusually sensitive to certain types of psychotropic medications due to reduced drug metabolism, reduced drug clearance and reduced plasma protein binding.
- Some underlying conditions such as Lewy body dementia may make the person particularly sensitive to the use of even small doses of a neuroleptic, e.g. haloperidol. For these reasons it is important to start medications at a lower dose, titrate slowly and review frequently (with a minimum of three-monthly reviews).
- Every effort should be made to keep the dose at the lowest level at which the drug is effective.
- Side effects may not always be reported by the service users due to impaired communication in many, and it is therefore essential for the prescribing clinician to make both the users and carers aware of what to look for in terms of side effects.

Use of antidepressants:

- To treat symptoms of depression Selective Serotonin Reuptake Inhibitors (SSRIs) are preferred but attention is needed to the risk of developing low sodium levels. Sertraline has been found to be effective in the treatment of depression in people with dementia in a randomised control trial (Lyketsos et al., 2003). There is emerging evidence (Nyth & Gottfries, 1990; Pollock et al., 2002,) that SSRIs may be used for treatment of agitation in dementia. Many clinicians prefer trazodone for which there is some evidence available for its effectiveness (Sultzer et al., 2002).

Use of mood stabilisers:

- Drugs such as carbamazepine or sodium valproate may be considered if there is evidence of rapid cycling mood disorder or significant mood fluctuations.

Key Points:

- Acetylcholinesterase inhibitors are used mainly in Alzheimer's type dementia. NICE guidance (2007) recommends their use in middle stage of dementia to delay the progress of the illness. However, NICE has restated that the difficulty in staging dementia in people with learning disabilities should not disadvantage them, thus introducing some flexibility for prescribers.
- Psychotropic medications have only a limited role in the management of neuropsychiatric symptoms in people with learning disabilities and dementia and should only be considered if other environmental/psychosocial approaches have produced only very limited or no benefit and the risk from the symptoms is assessed as high.
- Antidepressant medications are useful in the management of depressive illness in people with dementia and learning disabilities.
- Caution should be exercised in the use of antipsychotic medication in the context of the evidence of a high risk for cerebrovascular events and mortality.
- Where psychotropic medications are used, target symptoms should be clearly recorded; risks and benefits discussed with the person and/or carers and the minimum effective dose should be used for the shortest length of time.

Section 14 – End of life issues

Dementia is a progressive and, at present, terminal illness. Its course, whilst varying in detail from person to person, is by definition associated with the progressive loss of skills and the ability to communicate, ending in a final stage where the person becomes completely dependent on others for their every need. As the illness progresses, so it becomes harder for others to ascertain the wishes of the person suffering from dementia. For these reasons preparing for the end of life is of particular importance. Yet, compared to cancer care, there is limited attention paid to end of life and palliative care issues. However, the generally fairly long course of dementia allows time for the person him/herself and for others to prepare and to ascertain how they wish to be supported and to plan for how their health and well-being will be maintained.

End of Life issues are now beginning to be addressed through the Department of Health End of Life Strategy (DH, 2008), which sets out the key areas for action to ensure that people have excellence in end of life care. This includes developing strong links with services that provide excellence in end of life care, e.g. hospices, palliative care services, admiral nurses.

Good practice in dementia care requires the recognition that dementia is a progressive and terminal illness with a generally defined course. Thus, as new problems arise their occurrence should not be a surprise and should have been anticipated. In the early stages of dementia whilst the person may well be able to contribute, the issues listed below should be considered as these will all become relevant towards the end of the illness or at the person's death. To what extent this is possible will depend on the person's pre-existing level of disability, but in every case they should be considered within the context of the Mental Capacity Act.

- An understanding of where, how and by whom the person would like to be supported towards the end of their life and whether he/she would like to appoint someone to make healthcare decisions on his/her behalf when he/she no longer has the capacity to do so.
- Future management of financial affairs such as the appointment of a Lasting Power of Attorney and the making of a will.
- An understanding of the person's preference with respect to the use of or withholding of more invasive treatments in the advanced stages of dementia when he/she will lack the capacity to consent. These may include the use of artificial nutrition and hydration, ventilation, and the treatment of infections. If the person has the capacity to do so they may be supported to make an advanced statement with respect to these matters.

The nature of dementia is such that it is difficult to predict whether a person is reaching the end of their life and this can lead to difficulties in terms of access to palliative care services. However, NICE (2006) advises that a palliative care approach should be taken from the time of diagnosis until death. The aims are to support quality of life, to help the person die with dignity in a place of their choosing and to provide support to relatives and friends to help them prepare for the death. Such an approach considers the physical, psychological, social, cultural and spiritual needs of the individual.

The following are areas that may require particular attention towards the end of the person's life:

Eating and drinking: People should be supported and encouraged to eat and drink by mouth for as long as possible, with specialist advice being obtained concerning any feeding and swallowing difficulties from speech and language therapists, occupational therapists and/or dieticians. Weight loss is common in late-stage dementia. Nutritional support, including artificial (tube) feeding should be considered if dysphagia is thought to be a transient phenomenon. NICE do not recommend that such artificial means are used in those with severe dementia for whom dysphagia or disinclination to eat is a manifestation of dementia severity. Specific ethical and legal principles apply with respect to withholding or withdrawing nutritional support.

Resuscitation: It is generally considered that cardiopulmonary resuscitation is unlikely to succeed in cases of cardiopulmonary arrest in people with severe dementia. For those people who have retained their capacity to make a decision on this matter it is for them to decide whether or not to agree to resuscitation if it were needed. For those who now lack the capacity to make such decisions, any wishes expressed in a valid and applicable advanced decision to refuse treatment must be respected if the circumstances are applicable. The policies and procedures set out in the MCA (2005) (or similar legislation in other jurisdictions) should be followed.

Pain relief: Unexplained changes in behaviour or evidence of distress in a person with dementia may be indicative of underlying pain. The possible cause for pain needs to be investigated and necessary treatments undertaken and both pharmacological and non-pharmacological approaches to pain relief considered.

Posture: Management of posture is important in order to optimise remaining abilities and prevent the development and/or progression of secondary complications such as pain, fatigue, muscle shortening, joint deformity, respiratory complications and pressure ulcers. There needs to be assessment of postural management equipment needs such as standing aids (where appropriate), specialist seating provision and sleep systems. Good postural management can also facilitate safe eating and drinking and improve respiratory function.

Key points:

- Preparation for end of life should be ongoing as the dementia progresses.
- All care should be provided in accordance with the provisions of the Mental Capacity Act.
- Care includes both physical care and psychological care of the person, and psychological and emotional care of staff and carers.

Section 15 – Capable commissioning for people with learning disabilities and dementia

Most commissioners are still not aware of the increase in the prevalence of dementia amongst people with learning disabilities, despite the publication of the *Guidance for Commissioners* (Turk et al., 2001). With the move to social care commissioning, it is essential that commissioners are aware of both the health and social care needs of people who develop dementia. Health care commissioners will still have a responsibility for Continuing Healthcare (DH, 2007).

New funding arrangements give rise to a range of new options for funding care packages for people with dementia. However, there needs to be a recognition that dementia is by definition a deteriorating condition, and that peoples' needs will increase over time, and therefore more and timely funding will be needed to ensure safe practice.

Good quality care will involve active partnership-working between learning disability services, older people's services, primary and secondary health care, palliative care and social care. These areas should develop an integrated dementia strategy for the care of people with learning disabilities and dementia. This should involve the development of an integrated care pathway involving all relevant agencies. Specialist health professionals, e.g. speech and language therapists, physiotherapists, occupational therapists, dietitians and community learning disability nurses are all essential partners in providing excellence in care.

Staff involved in assessment and intervention need to be trained in dementia care and be able to offer both holistic and specialist assessments and a range of interventions aimed at meeting the needs of people with learning disabilities and dementia. Care managers have an essential role to play in ensuring that services are actively monitored to ensure that they are responsive to the changing needs of the person. People with learning disabilities and dementia should have access to regular reviews (three- to six-monthly depending on the rate of deterioration) by an identified care manager.

Terminal care needs to be planned for in advance, using the same principles and services available to the general population. Good partnership-working with palliative care services is essential, both to support the person and the carers.

Key points:

■ Each area should develop a dementia strategy and integrated care pathway to support the provision of high quality assessment, intervention and care for people with learning disabilities who develop dementia.

Section 16 – Capable workforce

The NICE guidance (2006) states that: *'Health and social care managers should ensure that all staff working with older people in the health, social care and voluntary sectors have access to dementia–care training (skill development) that is consistent with their roles and responsibilities'.* The need for effective training and leadership were also key issues in the CSCI (2008) report *'See me, not the dementia'.*

Carers, paid staff and professionals from all disciplines will require a basic awareness of dementia in order to provide effective care. There are many courses/workbooks available or being developed both locally and nationally which cover the main areas. Basic awareness training should cover as a minimum:

- What is dementia?
- Types and signs and symptoms of dementia.
- Working with people in a person-centred way.
- Using accessible information formats for person-centred plans and health action plans.
- Lifestory work.
- Use of medication in dementia.
- Communication skills.
- Roles of different health and social professionals.
- Safeguarding adults.
- Palliative care approaches.

Staff with specialist skills will need to be identified so that all the needs of people with learning disabilities and dementia can be met within the local service (refer to the interventions listed in Section 12). Some of the development may be able to be undertaken by professionals from local services. Highly specialist skills may be profession specific and/or need to be developed through completion of diploma or degree courses at Higher Education Institutions.

Some of the important specialist areas include expertise in the areas of:

- Assessments of physical and mental health, cognitive abilities, communication skills, sensory issues, risk and mental capacity.
- Management and maintenance of skills.
- Dementia Care Mapping.
- Management and maintenance of physical and mental health.
- Medication management.
- Epilepsy.
- Sleep management.
- Pain recognition and management.

- Communication.
- Eating and drinking.
- Dysphagia.
- Environmental adaptations.
- Mobility.
- Aids for daily living.
- Behavioural analysis and intervention.
- Falls prevention.
- Risk management.
- Reminiscence.
- Reality orientation.
- End of life issues.

Key points:

- Basic awareness training will be required for staff, professionals and carers.
- Staff with specialist skills need to be identified to ensure that people with learning disabilities and dementia have excellence in care.

Section 17 – References

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Good Practice Standards – Self Assessment checklist

This framework is designed to be used by members of Partnership Boards, or their equivalent and senior professionals (clinicians, social workers, managers, commissioners and those responsible for inspection and review) who have responsibility within a defined area or population for the provision of services to people with learning disabilities who develop, or at risk of developing dementia. It is a self-assessment checklist that should be used to establish the extent to which local processes and practices reflect 'best practice' as described in more detail elsewhere in this document.

The current provision and practices that are followed by services, should be reviewed against the standards that are described. Users, advocates and carers should also be involved in the review process. The standards apply to people living in family homes, health and social care provision (within the statutory and independent sectors). They also apply when out of area placements have been purchased by the relevant authorities.

The checklist should be:

- Reviewed in a multi-disciplinary/multi-agency setting, with the aim of achieving a consensus view about how local services compare against the Good Practice Standards.
- The team that carries out the review of standards should ensure that they have the appropriate membership to achieve a broad view of services.
- The team should include: Commissioners, practitioners from the Community Learning Disabilities Team, practitioners from the relevant Older Adults Mental Health Service, Social Services Care Managers, residential providers, day service providers, inspection teams, people with learning disabilities and carers.
- The reviewing team should refer to the relevant section in the guidance document to clarify the detail required to show compliance with the standard.
- Evidence for compliance with each standard should be demonstrated.
- A joint action plan should be developed to address any areas of need, or to build on current good practice.
- The action plan should be reviewed regularly to update on progress.

1. Legal framework and guidance

Standard		Green Amber Red	Action required	By whom	By when
1.	**People who develop, or are at risk of developing dementia have access to assessments and interventions that are delivered within current legislation and national guidance.** **Refer to Section 1.**				

Green: People who develop, or are at risk of developing dementia have services delivered within current legal and best practice requirements. These will include:

- Clear health and social care pathways that incorporate assessment of capacity, in line with the Mental Capacity Act.
- The presence of a local advisory group with clear terms of reference, to consider issues of best interest for people who lack capacity.
- Local care pathways are in place to ensure compliance with NICE/SCIE clinical guidelines on dementia and the proposed National Dementia Strategy.
- All people with learning disabilities who develop (or are at risk of developing) dementia have a person-centred plan.
- Health action plans are offered to all people with learning disabilities who are at risk of developing dementia.
- People with learning disabilities who are at risk of developing dementia have the opportunity to plan their future care and have considered advance decisions and end of life planning as part of their care package.
- There is a published local Safeguarding Adults policy that people with learning disabilities, clinicians and carers can access.
- Where people have 'out of area' placements purchased for them, the purchasing authority regularly checks that the provider also achieves these standards.

Amber: There are a few elements that require action in order to meet national guidance and to comply with current legislation.

Red: There are significant gaps between national guidance/legislation and local practice.

2. Population

<table>
<tr><th colspan="2">Standard</th><th>Green
Amber
Red</th><th>Action required</th><th>By whom</th><th>By when</th></tr>
<tr><td>2.</td><td>Each area has a register / database of people with learning disabilities, which identifies people with Down's Syndrome that can be used to plan and deliver effective services.
Refer to Section 2.</td><td></td><td></td><td></td><td></td></tr>
<tr><td colspan="6">Green: There is a regularly updated register / database of all adults with learning disabilities known to both health and social care in the catchment area. This includes the identification of people with Down's syndrome, and people who are the responsibility of the area but have services purchased for them in other areas.</td></tr>
<tr><td colspan="6">Amber: There is only a partial database, or it is not regularly updated, or it does not identify people with Down's syndrome, or it only identifies people with Down's syndrome, or excludes people who are placed by the authority in out if area accommodation.</td></tr>
<tr><td colspan="6">Red: There is no register / database or it is not regularly updated.</td></tr>
</table>

3. Multi-agency dementia strategy

Standard		Green Amber Red	Action required	By whom	By when
3.	**Each geographical area has an agreed multi-agency learning disabilities and dementia strategy for people with learning disabilities with an agreed action plan that addresses the standards outlined in this document.** **Refer to Section 15.**				
Green: There is an agreed comprehensive multi-agency learning disabilities and dementia strategy that addresses all of the standards that are detailed in this guidance document.					
Amber: There is either: only a single agency learning disabilities and dementia strategy, or: a multi-agency strategy for some aspects of the standards that are detailed in this document, but it is not comprehensive in scope.					
Red: There is no agreed strategy within any agency.					

4. Care Pathway

Standard		Green Amber Red	Action required	By whom	By when
4.	**People who develop, or are at risk of developing dementia have assessment, diagnosis, treatment and support delivered according to an agreed multi-agency care pathway which includes access to other services beyond the specialist learning disability areas (e.g. Older People's Mental Health Services, neurology, advocacy).** **Refer to Section 15.**				

Green: There is a multi-agency care pathway for assessment, diagnosis, treatment and support of people who develop, or are at risk of developing dementia, which has been agreed by the Partnership Board or its equivalent.

Amber: There is a single service care pathway, or only a partial care pathway or it is not fully agreed.

Red: No care pathway has been agreed.

5. Multidisciplinary approach to assessment, diagnosis and support

Standard		Green Amber Red	Action required	By whom	By when
5.	**People who develop, or are at risk of developing dementia are offered assessments, diagnosis and specialist support from the full range of dedicated and skilled professionals within learning disabilities services. They have easy access to specialist assessments within the general health services (e.g. neurology)** **Refer to Section 15.**				
Green: There is a full range of suitably trained professionals within the learning disability specialist services that fully matches the care pathway. They have easy access to specialist input from mainstream services such as neurology.					
Amber: The workforce only partially matches the care pathway.					
Red: There are significant gaps in staffing to deliver the care pathway.					

6. Assessment and diagnosis

Standard		Green Amber Red	Action required	By whom	By when
6.	**People who develop, or are at risk of developing dementia have easy access to comprehensive assessment and diagnostic services according to an agreed protocol.** **Refer to Sections 3 – 7.**				

Green: Within each area, people who develop, or are at risk of develop dementia have easy access to comprehensive assessment and diagnostic services where:

- Assessments are undertaken according to an agreed assessment protocol that includes decisions on whether the service offers baselines, reactive screening and / or prospective screening.
- Assessments follow an agreed assessment battery including both direct assessment of the person with learning disabilities and informant assessment.
- Assessments include physical, psychological, social and environmental factors.
- There is a timely response to referrals.
- Diagnosis is multi-disciplinary and based on established diagnostic criteria.
- There is a written formulation.
- There is a protocol for how to share the diagnosis of dementia both with people with learning disabilities and carers.
- Other illnesses / conditions are identified and treated promptly.

Amber: There are a few gaps that require action in order to achieve a comprehensive assessment and diagnosis service.

Red: There are significant gaps between the best practice standards for assessment and diagnosis, and the current local procedures.

7. Person Centred Dementia Care

Standard		Green Amber Red	Action required	By whom	By when
7.	**People who develop, or are at risk of developing dementia have their care provided according to person centred principles and this is individualised to meet the needs of the person.** **Refer to Section 10.**				

Green: People who develop, or are at risk of developing dementia have:

- A person centred plan that has been drawn up with the person to include future needs.
- Dementia care provided according to an agreed philosophy of care that places the person at the centre of the process.
- Care provided following the principles of the Mental Capacity Act and least restrictive practices.
- Care able to meet their changing needs without risk or delay.
- Care provided that promotes social inclusion.

Amber: Most people will receive care that meets this standard, but there are some gaps for a few people.

Red: Fewer than half of the people who have dementia or are at risk of developing dementia receive care that meets this standard.

8. Care management and review

Standard		Green Amber Red	Action required	By whom	By when
8.	**People who develop, or are at risk of developing dementia have their care purchased, monitored and reviewed by an effective care management system, whether in the geographical area or in an out of area placement.** **Refer to Section 15.**				

Green: There is a clear inter-agency care co-ordination system that ensures that all people with learning disabilities and dementia have:

- A named care manager.
- A written multi-disciplinary care plan incorporating the person's person centred plan.
- Regular care reviews.
- A health action plan that identifies pain signals and management.
- Proactive risk assessment and management plans in place.
- Access to flexible funding to meet changing needs without delay.
- Outcomes monitored regarding their quality of life.
- Staffing levels increased as required including the provision of waking night staff.
- Care that recognises that people with learning disabilities and dementia should not be moved unless absolutely necessary.
- No multiple moves.

Amber: Most elements of a comprehensive care management system are in place but there are a few gaps.

Red: There are significant gaps in the system.

9. Interventions

Standard		Green Amber Red	Action required	By whom	By when
9.	**People who develop dementia have prompt access to the full range of medical, psychological, therapeutic and social interventions as required.** **Refer to Sections 12 & 13.**				

Green: People who develop dementia have, as required, prompt access to staff who deliver:

- Medications in line with NICE guidance, according to a locally agreed protocol.
- Strategies, equipment and aids to maintain mobility, promote exercise, address posture, correct gait and reduce the risk of falls.
- Specialist pressure area care.
- Strategies for maintenance of adequate oral intake in a safe manner. This includes swallowing assessment, eating programme with dietetic advice along with advice regarding posture, and diet to reduce risk of constipation.
- Aids / adaptations to help to maintain continence.
- Strategies to aid communication including communication passports, objects of reference, pictures, signposting, signs and symbols.
- Additional supports to maintain self care and domestic skills and adaptation of the environment to aid daily living.
- Support for people to engage in failure free activities including sensory stimulation, aromatherapy, music and art therapy.
- Reminiscence, reality orientation, life story books, validation therapy.
- Dementia care mapping.
- Behaviour management including promotion of positive behaviour and feelings of self esteem, anxiety management, functional analysis and setting up of programmes for challenging behaviours, setting up of crisis intervention plans.

Amber: Most elements of a comprehensive intervention service are in place, but there are a few gaps.

Red: There are significant gaps in the range of services that are required.

10. Dementia friendly environments

Standard		Green Amber Red	Action required	By whom	By when
10.	**People who develop, or are at risk of developing dementia, have accommodation and day and leisure activities which are dementia friendly and are commissioned to meet their changing needs.** **Refer to Section 11.**				

Green: Most people in the area, who develop or are at risk of developing dementia, have accommodation and day and leisure activities which are dementia friendly and can meet their changing needs. Any accommodation and / or day and leisure activity will be appropriate in terms of:

- Have been identified as being ‘dementia friendly’.
- Have ensured that environments are safe, suitably stimulating, make sense and are predictable.
- Have ensured appropriate use of colour in homes and services.
- Have ensured appropriate furnishings are provided.
- Provide appropriate aids and adaptations in a timely manner.
- Provide appropriate wheelchairs, special beds and special seating in a timely manner.
- Use appropriate assistive technology.

Amber: Some people in the area will have access to accommodation and activities that meet these quality standards.

Red: Few people in the area have access to accommodation and activities that meet these quality standards.

11. 'Dying in place'

Standard		Green Amber Red	Action required	By whom	By when
11.	**People who develop dementia are supported to ‘die in place’, with additional supports provided in a timely manner.** **Refer to Section 11.**				

Green: There are processes in place to ensure that people who develop dementia are supported in ways that help to maintain them in their own home with:

- Good advance planning
- Additional staff as required including waking night staff
- Environmental adaptations as necessary
- A system for rapid availability of funding to meet changing needs
- Good end of life care

If the person needs to move, then the new service should be:

- Close to the person’s existing home
- Within learning disabilities services
- Allow links to existing day and leisure services to be maintained
- A final move

Amber: Most people will receive care that meets this standard, but there are some gaps for a few people, such that they are moved out of the learning disability services

Red: Fewer than half of the people who have dementia receive care that meets this standard, or people experience significant delays in providing necessary supports, or people are placed in nursing homes.

12. Choices and rights of people with learning disabilities and dementia

Standard		Green Amber Red	Action required	By whom	By when
12.	**The wishes, choices and rights of the person with learning disabilities who develops, or is at risk of developing dementia are respected, and this is evidenced in the care provided.** **Refer to Section 10.**				

Green: Within the identified area, people with learning disabilities who develop, or are at risk of developing dementia all have:

- A regularly updated Person Centred Plan.
- A regularly updated Health Action Plan.
- An individualised care plan that is developed with all services that the person utilises, and is coordinated across services.
- Accessible reports about their care.
- Accessible information about dementia.
- Support for their peers with regard to dementia.
- Appropriate advocacy to meet their needs.

Amber: There are some gaps in services so that most, but not all people have access to all these provisions.

Red: There are significant gaps, such that few people have access to all these provisions.

13. Support to family carers

Standard		Green Amber Red	Action required	By whom	By when
13.	**Family carers are assisted to understand and support their relative who has dementia. Their needs as carers are met.** **Refer to Section 10.**				

Green: Within the identified area, family carers are assisted to understand and support their relative who has dementia:

- All carers of a person with learning disabilities and dementia are offered a carer's assessment in line with Local Authority guidance.
- Respite care / short breaks are available to those with dementia who are living with a family member. The respite provision is able to provide 'dementia friendly' accommodation and services.
- Family carers are offered accessible information including: dementia in people with learning disability/Down Syndrome, thyroid disorder, depression, pain awareness, etc, as relevant to the individual person.
- Family carers are provided with support from the local Learning Disabilities Community Team as required to manage, for example, challenging behaviour, depression, epilepsy, eating and drinking, aids and adaptations.
- Family carers are offered training sessions to increase awareness of dementia and its management in the home setting.
- Family carers are consulted about the development of services to people with learning disabilities in their area.

Amber: Carers have access to most of these services and there are only a few gaps in such provision.

Red: There are significant gaps in the provision for carers.

14. Capable workforce

Standard		Green Amber Red	Action required	By whom	By when
14.	**People who develop, or are at risk of developing dementia, and their families, receive support, advice and care from a capable workforce that is appropriately skilled.** **Refer to Section 16.**				

Green: Within the identified area:

- Members of the Learning Disabilities Community Team are up to date in their knowledge and skills through continuing professional development opportunities with regard to: the risks of dementia, its early symptoms and progress and the methods of managing it. As a consequence, they can provide evidence-based assessment and advice to individuals and care teams.
- A rolling programme of dementia training is provided to the appropriate care staff in the identified area throughout the year. A log is kept of attendance. Homes caring for someone with dementia and learning disabilities are specifically targeted.
- Learning Disabilities Community Team staff provide training and support to care staff teams in residential, respite and day activity settings concerning the management of any individual with dementia, as required.
- There is a workforce development plan that addresses the issues of dementia care.

Amber: There are some gaps in the workforce development, with some staff not being suitably trained.

Red: There are significant gaps in the workforce development, with many staff not having the necessary skills to support people who have dementia.

15. End of Life Care

Standard		Green Amber Red	Action required	By whom	By when
15.	**People with learning disabilities and dementia have End of Life care delivered in line with the national strategy.** **Refer to Section 14.**				

Green: Within the identified area, End of Life care includes all the following elements:

- The person with learning disabilities and dementia has been involved as fully as possible in decisions and preferences with regard to end of life care.
- Carers of the person with learning disabilities and dementia have been involved in end of life discussions.
- Mental capacity assessment has taken place with regard to specific end of life decisions.
- Where possible, the person with learning disabilities has been given the opportunity to make Advance decisions about their care.
- Where the person does not have capacity, decisions are made following Best Interests guidance.
- Palliative care services are able to meet the needs of people with learning disabilities and dementia.

Amber: There are a few gaps in the provision of a comprehensive end of life care for people with learning disabilities, but most of the elements are available.

Red: There are significant gaps in provision, such that few people receive comprehensive end of life care.

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