



Optimum
Workforce Leadership

Rarer Types of Dementia Training

Pack for Care Homes



supported by



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County Council**



Health Education England

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Introduction

This training pack has been designed to increase your knowledge on rarer types of dementia and has been designed to be a self-learning tool. Based upon the knowledge that you have gained, the training pack has adopted a blended approach in which to consolidate your learning. The pack enables you to evaluate your learning through questionnaire, case studies and competency based assessment. Each assessment should be completed with the training pack facilitator/supervisor. Both you and your training facilitator/supervisor should:

- use the Competence Assessment Tool to assess yourself and devise an action plan to meet your individual development needs
- provide evidence for renewal of your registration with the Nursing and Midwifery Council revalidation
- provide evidence of achievement for your personal development plan
- use your assessment results to focus on your development needs, prepare for supervision meetings and support your career development.

While completing the tool it is useful to use the following framework in order to gain the maximum benefit from the training pack.

Learning and Development Framework

Step 1 Review and assess

Your knowledge, skills and attitudes using the training pack compile your evidence to support your assessment.

Step 2 Identify and prioritise your learning and development needs

From your assessment results including any 360° feedback. Identify, plan and prioritise your overall learning and development needs with your facilitator/supervisor.

Step 3 Plan and action

Discuss suitable learning opportunities with your supervisor and agree relevant learning outcomes. Record these in your learning and development plan.

Step 4 Evaluate your learning and development

In relation to improvements in your knowledge, skills and attitudes. Maintain a reflective record of your learning and development in your portfolio, to support your preparation of your supervision sessions or development review meetings.



Rarer dementias

As well as the more common dementias there are also many other less common types. There are over a 100 different types of dementia and understanding some of the less common types of dementias is important. Subtle differences in presentation can help staff to understand resident's unique needs. It is not possible to list all types of dementias but for the purpose of this learning resource various types are incorporated which the author believes you may come across professionally as follows:

Frontotemporal dementia

The shrinkage of tissue in the frontal and temporal lobes of the brain that affects personality and behaviours is considered Frontotemporal dementia, or FTD.

According to the National Institute of Neurological Disorders and Stroke Pick's disease, Primary Progressive Aphasia, Behavioural variant Frontotemporal dementia and Semantic dementia all fall under the umbrella of Frontotemporal dementia.

Frontotemporal dementia accounts for approximately 10 percent of all dementia cases. FTD is also sub divided in to three sub types. General common symptoms are described as:

- language impairments ranging from speaking in broad terms with less meaning to having trouble forming words altogether
- involuntary movements
- lack of coordination
- loss of balance representing mobility issues experienced by those with frontotemporal dementia.

Sub type 1 Behavioural variant frontotemporal dementia (bv FTD)

The following symptom must be present to meet criteria for bv FTD

- Shows progressive deterioration of behaviour and/or cognition by observation or history (as provided by a knowledgeable informant).



Three of the following behavioural/cognitive symptoms (A–F) must be present to meet criteria. Ascertainment requires that symptoms are persistent or recurrent, rather than single or rare events.

- A** Early* behavioural disinhibition [one of the following symptoms (A.1–A.3) must be present]:
 - A.1** Socially inappropriate behaviour
 - A.2** Loss of manners or decorum
 - A.3** Impulsive, rash or careless actions.
- B** Early apathy or inertia and one of the following symptoms (B.1–B.2) must be present]:
 - B.1** Apathy
 - B.2** Inertia.
- C** Early loss of sympathy or empathy and one of the following symptoms (C.1–C.2) must be present]:
 - C.1** Diminished response to other people’s needs and feelings
 - C.2** Diminished social interest, interrelatedness or personal warmth.
- D** Early perseverative, stereotyped or compulsive/ritualistic behaviour and one of the following symptoms (D.1–D.3) must be present]:
 - D.1** Simple repetitive movements
 - D.2** Complex, compulsive or ritualistic behaviours
 - D.3** Stereotypy of speech.
- E** Hyperorality and dietary changes and one of the following symptoms (E.1–E.3) must be present]:
 - E.1** Altered food preferences
 - E.2** Binge eating, increased consumption of alcohol or cigarettes
 - E.3** Oral exploration or consumption of inedible objects.



F Neuropsychological profile: executive/generation deficits with relative sparing of memory and visuospatial functions all of the following symptoms (F.1–F.3) must be present]:

F.1 Deficits in executive tasks

F.2 Relative sparing of episodic memory

F.3 Relative sparing of visuospatial skills.

III. Probable bv FTD

All of the following symptoms (A–C) must be present to meet criteria.

A Meets criteria for possible bv FTD

B Exhibits significant functional decline (by caregiver report or as evidenced by Clinical Dementia Rating Scale or Functional Activities Questionnaire scores)

C Imaging results consistent with bv FTD one of the following (C.1–C.2) must be present]:

C.1 Frontal and/or anterior temporal atrophy on MRI or CT

C.2 Frontal and/or anterior temporal hypoperfusion or hypometabolism on PET or SPECT scan.

IV. Behavioural variant FTD with definite FTLD Pathology

Criterion A **and either** criterion B or C must be present to meet criteria

A Meets criteria for possible or probable bv FTD

B Histopathological evidence of FTLD on biopsy or at post-mortem

C Presence of a known pathogenic mutation.



Exclusionary criteria for bv FTD

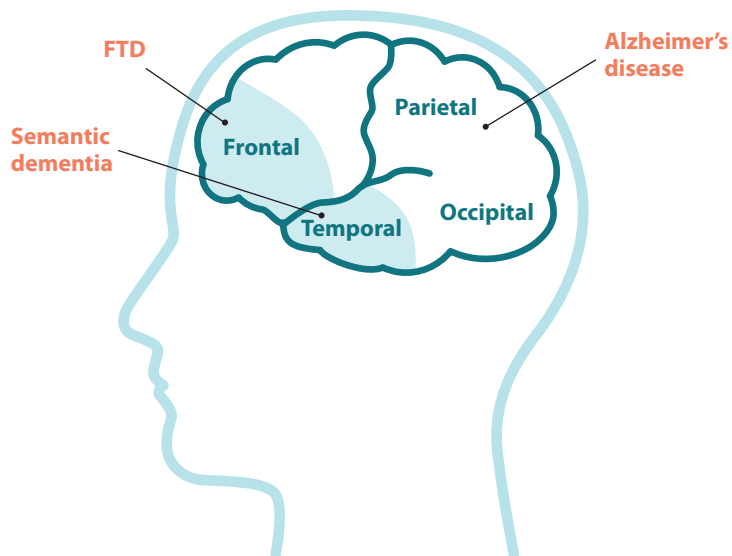
Criteria A and B must be answered negatively for any bv FTD diagnosis. Criterion C can be positive for possible bv FTD but must be negative for probable bv FTD.

- A Pattern of deficits is better accounted for by other non-degenerative nervous system or medical disorders
- B Behavioural disturbance is better accounted for by a psychiatric diagnosis
- C Biomarkers strongly indicative of Alzheimer's disease or other neurodegenerative process.

***As a general guideline 'early' refers to symptom presentation within the first three years.

Making a correct diagnosis is not easy and is why many people with this type of dementia get misdiagnosed, undiagnosed or labelled as having behavioural problems.

Neuro imaging of areas of the brain predominately affected by Frontal temporal dementia and Semantic dementia:



References: Table reprinted from Brain; permission granted to AFTD from Copyright Clearance Center. Rascovsky K et al. *Sensitivity of revised diagnostic criteria for the behavioural variant of frontotemporal dementia*. Brain. 2011 Sep, 134(pt9): 2456-77. Epub 2011 Aug 2.



Symptoms and management

Meeting the needs of people with Frontotemporal dementia (FTD)

FTD is a condition associated with people under 65 years of age and is described as a cortical dementia. Symptoms may not manifest themselves for some time and carers usually portray a clinical picture in which change in personality is marked. Lack of understanding associated with this type of dementia often leads to both informal and formal carers struggling to understand people's behaviours. Frequently people with this type of dementia can easily be labelled as 'doing things on purpose' or are 'problematic'. This is usually because people struggle to understand that this is a type of dementia as memory is fairly well preserved initially.

Unlike other dementias initially memory is less affected and due to this staff often struggle with behaviours as they struggle to understand that this still remains to be a dementia. Social disinhibition, impulsivity all compound the problem. Patience and understanding is required while ensuring that resident's privacy and dignity is maintained. Someone with FTD may appear 'selfish' and lacking in patience in their quest to have something right now. They may have a constant need for sugary foods and demonstrate a lack of understanding or appreciation of other needs. Crucially people with FTD may demonstrate disinhibited behaviour making them unpopular with fellow residents and visitors.

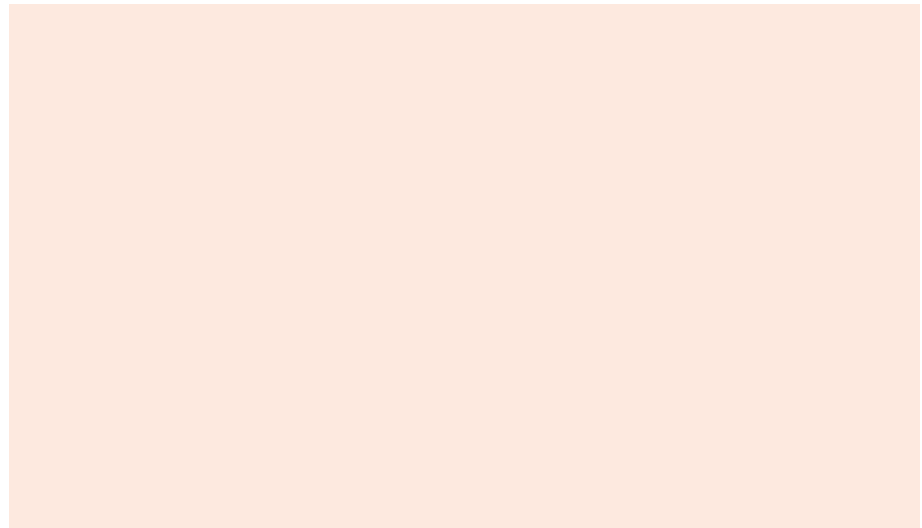
It is not easy but as much as possible preserving dignity while maintaining confidentiality is required. Some residents may display deep apathy while still sustaining the physical ability to complete the task you are asking them to complete appreciation of this is also important. The old saying 'where there's a will there is a way' does not apply!

Remember people with FTD do not have a stop and start button and this may manifest itself in repetitive behaviours. Frustration may be evident as they struggle with planning and therefore careful consideration is required before tasks are attempted or when being asked to engage in any activities. A non-judgemental approach is needed based on resident's impaired judgment and decision making!



Time Out

Do you have a resident with FTD? Take a few minutes to consider their behaviours and what you can do to help them and minimise the effects of this type of dementia.



Primary Progressive Aphasia (Sub type FTD)

What is Primary Progressive Aphasia (PPA)?

Primary Progressive Aphasia (PPA) is a neurological syndrome in which language capabilities become slowly and progressively impaired. Unlike other forms of aphasia that result from stroke or brain injury, PPA is caused by neurodegenerative diseases, such as Alzheimer's disease or frontotemporal lobar degeneration.

PPA results from deterioration of brain tissue important for speech and language. Although the first symptoms are problems with speech and language, other problems associated with the underlying disease such as memory loss, often occur later. PPA commonly begins as a subtle disorder of language, progressing to a nearly total inability to speak, in its most severe stage.



The type or pattern of the language deficit may differ from person to person. The initial language disturbance may be fluent aphasia (the person may have normal or even increased rate of word production) or non-fluent aphasia (speech becomes effortful and the person produces fewer words).

A less common variety begins with impaired word-finding and progressive deterioration of naming and comprehension, with relatively preserved articulation.

As with aphasia that results from stroke or brain trauma, the manifestations of PPA depend on what parts of the left hemisphere are relatively more damaged at any given point in the illness. The person may or may not have difficulty understanding speech. Eventually, almost all people become mute and unable to understand spoken or written language, even if their behaviour seems otherwise normal.

Signs and symptoms of other clinical syndromes are not found through tests used to determine the presence of other conditions. PPA is not Alzheimer's disease. Most people with PPA maintain ability to take care of themselves, pursue hobbies, and, in some instances, remain employed.

Primary progressive aphasia (PPA) is the FTD subtype characterized by progressive loss of oral and written language skills as stated. Comprehension and language expression may be involved. When the problem is primarily with anomia and loss of word meaning, it is referred to as the semantic variant of PPA. Here, the meaning of specific words is lost, and both comprehension of the word and the ability to retrieve the name of an object may be lost. Speech remains fluent and grammar is good, however, paraphasias (word substitution errors) are common. This subtype of PPA is usually associated with TDP-43 pathology.

On the other end of the spectrum, speech production may be the primary symptom, and the term nonfluent/agrammatic variant of PPA is used. In this case, individuals lose grammar (the small connecting words) but have preserved language comprehension for specific items/objects. This causes speech to become effortful, hesitant, and sentence length becomes progressively truncated. Writing and language comprehension may be affected in the same manner. This subtype of PPA is usually associated with tau pathology. Logopenic PPA is the third major PPA variant. In these individuals, speech is slow, but grammar and comprehension are less affected. Impairment in the repetition of multisyllabic words and particularly phrases is a key feature. Sound substitution (phonemic) paraphasias are also seen in this group, as in a false word that rhymes with the intended word. Logopenic aphasia is usually associated with an underlying Alzheimer's pathology.



Treatment or assistance for residents with PPA

People with primary progressive aphasia are fighting against a condition in which they will continue to lose their ability to speak, read, write, and/or understand what they hear. Usually people with aphasia that results from stroke or head injury will experience improvement over time, often aided by speech therapy. This is not the case for people with primary progressive aphasia. However, individuals with PPA may benefit during the course of their illness by acquiring new communication strategies from speech-language pathologists. Some families have also learned new strategies with the support of Speech and Language therapy

Many people with aphasia find it helpful to carry identification cards and other materials that can help explain the person's condition to others. ID cards are available from the National Aphasia Association website.

Some communication-assistive devices may also be helpful. Non-verbal techniques for communicating, such as gesturing and pointing to pictures, may help people with PPA to express themselves.

Primary progressive aphasia (PPA) can leave affected persons and their loved ones with many questions and a sense of helplessness about the decline in the ability to communicate verbally and to read and write. These feelings may have been present for some time, but once the diagnosis of PPA is made, the questions tend to shift from "What is this problem?" to "What now? What can be done about this?"

By definition, PPA is progressive, but it is natural to ask what can be done to improve or maintain the ability to understand and express language. These questions cannot yet be answered with certainty. Although hundreds of cases have now been reported in the literature and thousands of people are probably affected, PPA is considered a rare condition. In addition, it may have more than a single underlying cause and there are few reports about the effectiveness of treatment for it. Nonetheless, it appears that some steps can be taken to help manage the communication problems.

The following overview offers some basic guidelines expressed in general terms. Because the severity and nature of the communication difficulties vary greatly, any treatment must be tailored to the particular individual needs and interests.



The medical perspective regarding treatment options

Most approaches to treating aphasia can be categorized as either medical or behavioural. From the medical perspective, there are currently no drugs or other interventions specifically designed for PPA. This partly reflects our limited understanding of what causes PPA and the likelihood that it has more than a single cause.

Neurologists sometimes prescribe drugs that are used for people with Alzheimer's disease under the assumption that PPA and Alzheimer's disease may share a common cause.

This assumption is unlikely to be true in the majority of cases. Currently, there are only anecdotal reports that the same drugs that target Alzheimer's disease are helpful in relieving the signs and symptoms of PPA.

The behavioural perspective regarding treatment

The primary approaches to managing PPA at this time are behavioural. That is, there are things that the person with PPA can do that may lessen the impact of the disease. Behavioural approaches emphasise practice, and counselling to enhance the ability to communicate, or compensate for the inability to communicate in conventional ways.

Some behavioural approaches for PPA are directed at improving or maintaining (in the short term) impaired language abilities. The decision to pursue this type of therapy should take into account the following considerations:

- the person with PPA must still have some capacity for insight, motivation and learning. Without them the possibility of meaningful improvement is greatly reduced
- the individual's significant others must be motivated and involved as well
- they play an important role in working on practice activities beyond formal therapy sessions and in providing cues for using effective communication strategies
- everyone involved must understand that therapy will not eliminate difficulties with communication. Even if there is improvement in communication ability, it will not reverse the progression of the disease.



The skills targeted for therapy are generally based on three factors: those abilities that are declining, those that may be relatively preserved, and those that are most important to the affected person. In all instances, therapy requires work that might be called "focused exercise of the brains language system.

At this time a small number of studies - all of which are based on only one person or a few carefully selected people with PPA - have documented improvements in abilities targeted by therapy. Skills that have improved include:

- comprehension of spoken instructions
- questions, production of sentences
- retrieval of words
- number reading.

It is premature to conclude that such treatments are likely to be effective for many people with PPA. However, these studies do suggest that for some affected individuals, and for some deficits, therapy may be beneficial. Whether these benefits continue beyond the period of formal therapy is not known.

Compensatory strategies for both residents and carers

Other behavioural approaches emphasise compensatory strategies that can improve communication, although not necessarily in conventional ways. Compensatory strategies can be 'resident-oriented' or 'other-oriented,' or a combination of both. Resident oriented strategies reflect things that the person with PPA can do to enhance communication, such as establishing the topic at the outset of a conversation, using gestures, and using pictures, writing or drawing (if still capable).

'Other-oriented' strategies include paying full attention to the affected person, giving feedback about the need for clarification, providing more time for communication, confirming information, keeping statements relatively brief, and supplementing speech with gestures.

Joint efforts on the part of both the person with PPA and others include speaking in environments that are conducive to effective communication (such as face-to-face conversations with minimal noise and other distractions).



People with PPA and their significant others often benefit from following the rule that communicating requires everyone's full attention. The notion that people with PPA require all the fuel in their 'language tank' when communicating is a useful analogy and one to keep in mind when engaged in speaking, listening, reading or writing activities.

It can be very helpful to consult with a speech-language therapist (SLT) to identify important communication needs, learn how and when compensatory strategies can best be used, and practice their use. The SLT can help to identify specific strategies and investigate whether augmentative strategies (for example, gesture, pantomime, and drawing) may supplement or sometimes replace verbal communication. Such strategies have been reported as helpful in some people with PPA.

Electronic/computer devices may be able to supplement or replace speech in some people with PPA. Because they require a person to use conventional language or other symbols, they may not be helpful for those whose language skills are already severely impaired. Some people with PPA also have an apraxia of speech (AOS), a problem with the programming of movements for speech rather than a language problem). They may have speech that is far more impaired than their language comprehension or ability to read and write. As long as their ability to control movements of body parts needed to use the devices is relatively intact, those with AOS and relatively mild PPA may be good candidates for electronic/computer alternatives to speech.

Generally, the development, practice and learning of augmentative or alternative means of communication should occur well before there is an actual need to use them, so they are readily available and more easily used when and if the need emerges.

Finally, there can be little doubt that simply learning about PPA is beneficial, both psychologically and practically. As discussed above, SLTs who have experience working with people with aphasia and degenerative neurological diseases can address questions about aphasia in general, and PPA in particular, and can help plan for future communication needs.

Some PPA residents and/or spouses benefit from joining an Aphasia Community Group or Stroke Support Group that has others with aphasia in it. This is true even if the others do not have aphasia that is progressive.

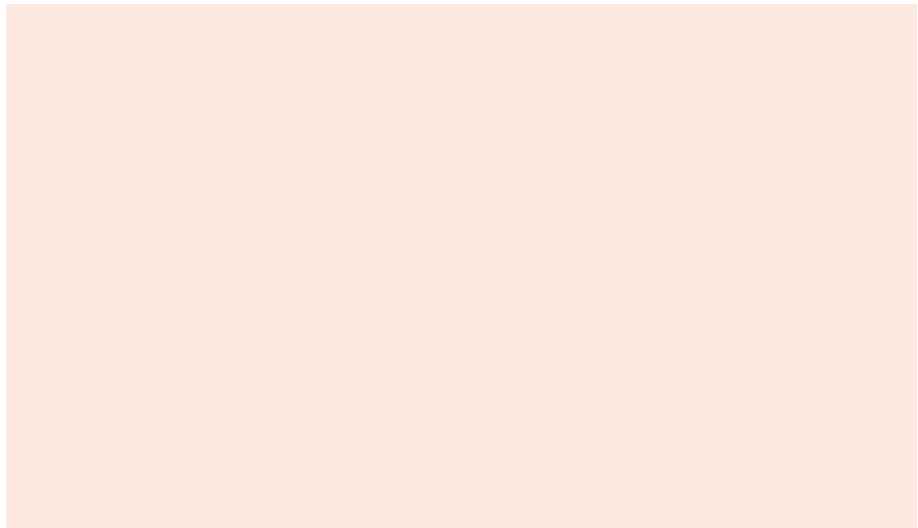


The National Aphasia Association (NAA) provides information about PPA and listings of support groups (www.aphasia.org).

Viewed in the most positive sense, the diagnosis of PPA does not mean the end of communication. It can be the first step to identifying ways to maintain communication abilities for as long as possible.

Time Out

Based on the information above can you consider how you and other professionals may be able to support the needs of residents with Primary Progressive Aphasia as independently as possible.



Semantic dementia

Semantic dementia (SD), is a progressive neurodegenerative disorder affecting language. Semantic dementia (SD) is described as progressive and relatively circumscribed loss of semantic knowledge, and falls under the broader umbrella of frontotemporal dementia, and was officially identified as a clinical syndrome less than 50 years ago.

From a neuroimaging perspective, SD is characterised by hallmark asymmetrical atrophy of the anterior temporal pole and anterior fusiform gyrus, which is usually left lateralised. Functional magnetic resonance imaging (fMRI) studies have revealed widespread changes in connectivity with relative preservation of frontal and parietal regions alongside preserved memory performance.

Recent research has demonstrated strong clinicopathological concordance in SD, with TDP43 type C as the most common pathological subtype.

Moreover, an underlying genetic cause appears to be relatively rare in SD, with the majority of cases having a sporadic form of the disease.

The relatively clear diagnosis, clinical course, and pathological homogeneity of SD make this syndrome a promising target for novel disease-modifying interventions. The development of neuroimaging markers of disease progression in future may help.

Clinically, residents with SD show a speech profile that is relatively fluent but empty of content, producing a pattern of so-called logorrhoea. Impaired word comprehension is a mandatory feature and residents demonstrate word alienation in that they are able to repeat words such as “violin” or “caterpillar” but have no idea of their meaning.

This deficit gradually progresses from low frequency and less familiar words, such as those mentioned, to more common words. SD residents are also impaired on nonverbal semantic matching tasks, tests of colour knowledge, sound knowledge, and object-use knowledge, which do not require naming or verbal comprehension even from an early stage of the disease.

Such findings have provided evidence that, in SD, symptomatology reflects a profound and progressive loss of conceptual knowledge which is not limited to performance on verbal tasks.



There is also accompanying surface dyslexia: residents are unable to correctly pronounce irregular words such as “pint” which they read to rhyme with “hint or flint”.

In contrast, recent studies have confirmed that episodic memory is relatively preserved in SD. Residents typically show relatively preserved recollection of recent autobiographical memory in the context of poorer remote autobiographical memory.

Changes in behaviour and social cognition are increasingly recognised in SD. Clinically, SD residents often show mental rigidity and inflexible behaviour. For example, residents may become obsessive in tasks they engage in (e.g. we have noticed residents spending hours completing jigsaw puzzles), food preferences (usually restricted to specific foods), or daily routines (e.g. clockwatching).

In addition, SD residents may have increased apathy and changes in eating behaviour, as well as loss of empathy, impaired emotion perception and emotional memories, and reduced theory of mind capacity.

Over time, many residents become essentially mute with only a limited repertoire of stereotypic phrases and a complete loss of word comprehension. Changes in emotional capacity as well as increased rigid behaviours are associated with higher carer burden and progressive behavioural changes and/or increasing disability leads to residential care in most cases.

Cognitive profile of person with semantic dementia at presentation:

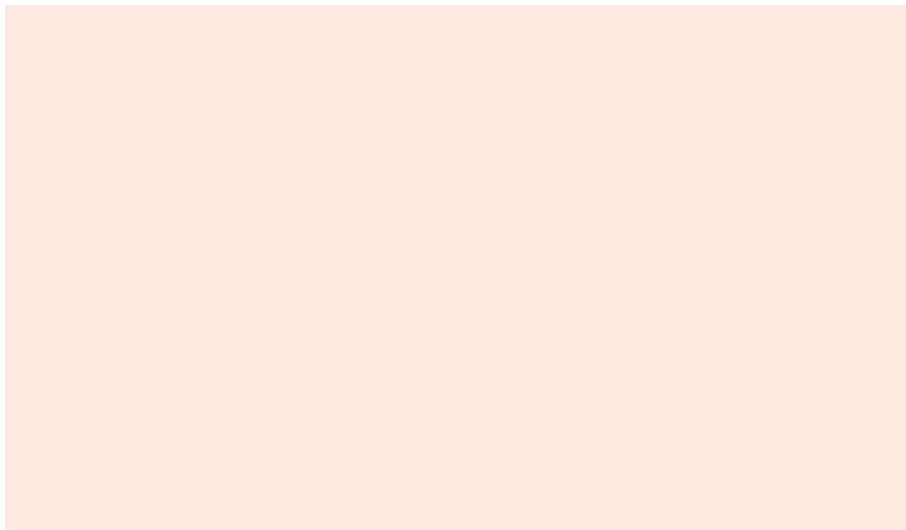
Impaired	Relatively preserved
Confrontation naming	Episodic memory
Word comprehension	Navigation
Object recognition	Visuospatial
Autobiographical memory	Attention
Future thinking	Processing speed
Emotion perception and apathy	Phonology and syntax
Theory of mind	Nonverbal problem solving



SD appears to be one of the more straightforward frontotemporal dementia subtypes. It has a clear clinical course, which begins with language features and, with progression, affects behaviour and social cognition; this reflects early and relatively circumscribed neurodegeneration of the anterior temporal pole, which encroaches into medial prefrontal and posterior temporal regions as well as into the contralateral hemisphere with disease progression.

Time Out

Based upon semantic dementia presentation consider some meaningful activities that could be organised to support a resident who may have SD?



FTD and motor neurone disease (MND)

As many as 20% of FTD people develop signs of (MND). Likewise approximately half of those with amyotrophic lateral sclerosis (ALS) develop some fronto-executive problems. A smaller number of these people will develop full-blown FTD-ALS. Many people that have the combination syndrome or gene, and thus a hereditary form of the disease.

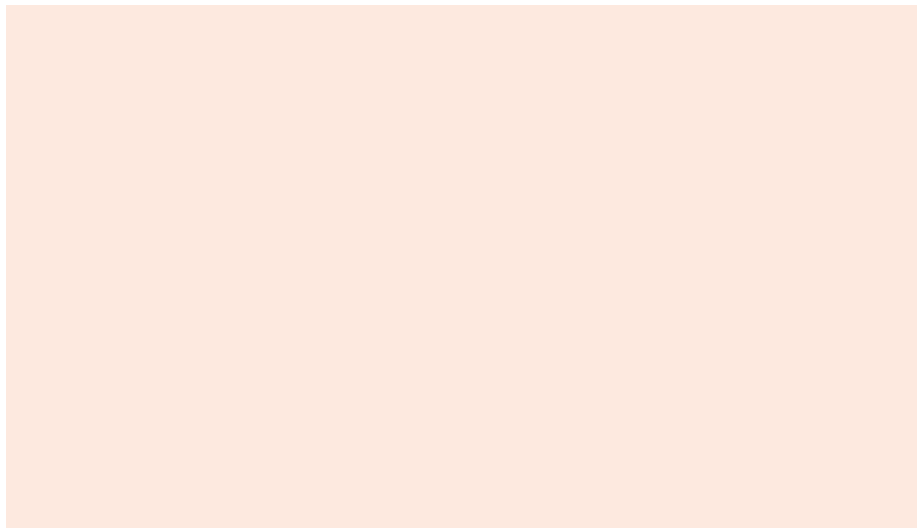


With or without the gene expansion, the addition of motor neuron disease to FTD is a compromising factor which greatly reduces their median survival rate to approximately less than three years. Behavioural symptoms complicate the management of dysphagia as well as respiratory dysfunction as respiration therapy and percutaneous endoscopic gastrostomy (PEG) feeding tubes are not well tolerated by the person. Evidence has shown PEG feeding shows little survival benefits.

Although significant phenotypic heterogeneity exists among C9orf72 carriers, the majority present with either bvFTD or FTD-ALS. PPA variants including nonfluent/agrammatic and semantic are rare. Prominent psychosis with delusions and hallucinations are relatively common (Boeve et.al 2012). Research into the clinical features associated with the C9orf72 mutation is active and on-going.

Time Out

Consider how you can support the needs of residents with a diagnosis of FTD and MND. Consider their long term planning arrangements and needs.



Other variants

FTD subtypes may also be associated with the Parkinson's plus syndromes of progressive supranuclear palsy (PSP) or corticobasal syndrome (CBS). CBS-like symptoms of asymmetrical movement problems, abnormal muscle tone, complex tremors, myoclonus and limb apraxia form the core CBS phenotype.

A recently appreciated speech/language variant known as primary progressive apraxia of speech (PPAOS) tends to precede the evolution into the nonfluent/agrammatic subtype; these patients experience articulatory difficulties but are not truly aphasic.

Creutzfeldt Jakob Disease (CJD)

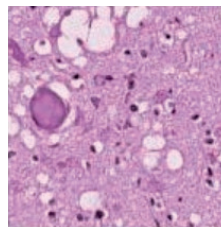
(CJD) (previously known as mad cow disease)

Creutzfeldt-Jakob disease, or CJD, is a degenerative brain disorder that presents as rapidly advancing dementia followed by death, according to the National Institute of Neurological Disorders and Stroke, or NINDS. Nine out of 10 of those afflicted with the rare disease die within 12 months from onset.

Brain shrinkage and deterioration occurs rapidly



© A.D.A.M



Brain section showing spongiform pathology characteristics of Creutzfeldt-Jakob



Symptoms of CJD include impaired judgment, lack of coordination, involuntary movements, depression and blindness, Scientists believe that a type of protein called prion in its infectious form, or a mutation of its genes, may be the cause of CJD. As of 2016, there is no cure, treatment or method of controlling the advance of CJD once it starts. Changes in your loved one's mood and temperament are often the early signs of Creutzfeldt-Jakob disease (CJD). They can also be particularly tough for the caregiver to deal with as you forget that it is not personal, it's the disease changing them. A resident with CJD will display many complex behaviours (but not all).

Note the following support that can be offered to a resident with CJD.

For anxiety and agitation

- See if you can figure out what triggers the anxious or agitated behaviour – you might be able to avoid setting off the behaviour by changing the environment or routine.
- Make sure the resident isn't in pain, hungry or thirsty or doesn't need to use the bathroom – they may be agitated because they can't express what they need.
- If they are aggressive or angry, don't touch them – it may be threatening instead of comforting.
- Talk about things that they love to help calm them.
- Speak in a calm voice and don't let their agitation agitate you.
- Try to make sure they are getting enough sleep
- Try not to rush or hurry them in anyway.
- Follow a regular daily schedule as much as possible.
- Eliminate stressful stimuli and maintain a quiet environment.



For depression and social withdrawal

- Try to keep them engaged with activities that suit his/her abilities – card games, reading aloud, talking.
- Physical exercise helps minimize depression – if their condition permits, walk, swim, garden or do something else they enjoy.
- Visitors might help improve mood, especially in the early stages, but watch for signs of agitation or fatigue.
- Validate and accept their feelings – don't try to force them to be happy and social they are intuitive.
- Talk to your doctor about medications that can help.

Abnormal beliefs (delusions)

Some people with CJD may have abnormal beliefs called delusions. These delusions can frighten them or cause unusual behaviour. Try to:

- avoid arguing or disagreeing with them by denying the delusion, try to be reassuring and comforting instead
- redirect from the delusion to a soothing activity
- turn off a television, radio, or computer if you think that is causing the delusion –they may not be able to tell the difference between entertainment and reality
- limit environmental activity and noise
- use a calm quiet approach and create a quiet environment
- keep areas well-lit so things in the room are not misidentified.



Abnormal sensory experiences (hallucinations)

Some people with CJD may see or hear things that no one else does or experience them differently than the others around them. It can frighten them. Try to:

- avoid arguing or disagreeing denying the hallucination, try to be reassuring and comforting instead.
- redirect them from the hallucination to a soothing activity
- limit environmental activity and noise
- use a calm quiet approach and create a quiet environment
- keep areas well lit so things in the room are not misidentified
- cover glass tables, mirrors and other pieces of furniture that have a high gloss and may create visual disturbances
- avoid the flickering, changing light of a TV which might prompt hallucinations
- turn off a television, radio, or computer if you think that is causing the hallucination again they may not be able to tell the difference between entertainment and reality.

Memory loss and confusion (dementia)

Most people with CJD experience progressive memory loss, confusion and impaired thinking, which can in turn add to frustration, distress and agitation. Try to:

- limit environmental activity and noise which may add to confusion
- follow a regular daily routine
- reminisce by telling stories from your shared past (“I remember when ...”)
- watch how visitors affect them and keep visits short if they cause distress
- keep to the same carer’s over time. New faces may cause anxiety.
- limit choices when you can – present two options for dinner or two options for clothing and let them choose. Too many choices can be overwhelming.



- minimize distractions like television and radio so that it is easier to focus on the task at hand
- speak to them with a calming tone and comfort them
- speak in shorter, simpler sentences
- see if cognitive activities like puzzles, block stacking, or picture matching help and interest them.

Balance and walking problems (ataxia)

With CJD, most people experience increasing coordination, balance and walking problems. Try to:

- remove obstacles that can cause a person to fall (throw rugs, toys, things around shin level)
- encourage the use of hand rails and/or grab bars where you can
- use a shower seat when possible or other aids
- make sure hallways and rooms are well lit – try plug-in night lights for low level lighting
- use lifts, standing frames, or gait belts to provide safe support.

Muscle jerks (myoclonus)

Some people with CJD may experience sudden, involuntary muscle jerks called myoclonus. A hiccup is a type of myoclonus. Generally, it doesn't hurt the person experiencing it and can be treated with medication. Try to:

- see if you can identify a specific noise, movement or experience that triggers the movements and then reduce or eliminate that trigger
- use a calm quiet approach and create a quiet environment
- minimise touching, turning and movement
- relax the muscles though aromatherapy, massage, physiotherapy etc.



Dance-like movements (chorea)

While chorea is a possible symptom, the irregular movements that flow from one muscle to another in a dance-like manner are not common in people with CJD. Try to:

- see if you can identify a specific noise, movement or experience that triggers the movements and then reduce or eliminate that trigger
- minimise touching, turning and movement
- talk to resident GP about available treatments.

Rigid posture (dystonia)

Dystonia is the movement disorder where sustained muscle contractions lead to stiffness and abnormal postures. Try to:

- massage etc
- use warm towels or a heating pad on the stiff muscles
- take deep, regular breaths to help relax if possible
- see if you can detect a trigger that causes the stiffness to start or worsen and then reduce or eliminate that trigger
- encourage exercise each day, if possible
- give your residents one some tonic water – it contains quinine, muscle relaxant
- talk to resident GP about medical options of causing distress.

Difficulty swallowing (dysphagia)

Some people with CJD may develop difficulty eating and swallowing (dysphagia) as their motor problems progress. Try to:

- obtain SALT assessment or DTN assessment
- discuss preparing pureed food or thickened liquids with DTN nurse or SLT professional



- use nutritional drinks to supplement a decreased diet (ask your health care team for suggestions)
- find out how to give regular mouth care, which can comfort them tremendously.
- let them if possible guide their food choices – their needs may alter as the disease progresses.

Loss of bladder control (urinary incontinence)

The loss of bladder control, incontinence, can cause agitation, restlessness and distress. Try to:

- encourage regular toileting routines dependent on level of independence.
- see if your loved one can practice double voiding (urinating, waiting a few seconds and then urinating again)
- discuss ways to address incontinence with resident GP / Community Matron/ Continence team.

Progressive blindness

Progressive blindness occurs in some CJD patients. Try to:

- ensure there is good lighting both inside and out
- remove excess clutter from rooms corridors etc
- get them a radio if they like to listen to stories or music.



Binswanger Disease

Binswanger's disease (BD), also called subcortical vascular dementia, or white matter disease, is a rare type of dementia caused by widespread areas of damage to the deep layers of white matter in the brain. The damage is the result of the hardening and narrowing (atherosclerosis) of arteries that supply the subcortical areas of the brain. As the arteries become more and more narrowed, the blood supplied by those arteries decreases and brain tissue dies. A characteristic pattern of BD-damaged brain tissue can be seen with brain imaging techniques such as CT scans or magnetic resonance imaging (MRI).

The symptoms associated with BD are related to the disruption of subcortical neural circuits that control executive cognitive functioning: short-term memory, organization, mood, attention, the ability to act or make decisions, and appropriate behaviour. The most characteristic feature of BD is psychomotor slowness - an increase in the length of time it takes for a thought to be translated into action, for example, for the fingers to turn the thought of a letter into the shape of a letter on a piece of paper. Other symptoms include forgetfulness (but not as severe as the forgetfulness of Alzheimer's disease), changes in speech, an unsteady gait, clumsiness or frequent falls, changes in personality or mood (most likely in the form of apathy, irritability, and depression), and urinary symptoms that aren't caused by urological disease.

Diagnosis

Brain imaging, which reveals the characteristic brain lesions of BD, is essential for a positive diagnosis.

Treatment

There is no specific course of treatment for BD. Treatment is basically symptomatic. Antidepressant medications such as Serotonin-Specific Reuptake Inhibitors (SSRI) such as Sertraline, or Citalopram are indicated for people with depression or anxiety. Atypical antipsychotic drugs, such as Risperidone and Olanzapine, may be helpful for individuals with agitation and disruptive behaviour. Recent drug trials with the drug Memantine have shown improved cognition and stabilization of global functioning and behaviour. The successful management of high blood pressure and diabetes can slow the progression of atherosclerosis,



and subsequently slow the progress of BD. Since there is no cure, the best approach is preventive, early in the adult years, by controlling risk factors such as hypertension, diabetes, and smoking.

Prognosis

BD is a progressive disease; there is no cure. Symptoms may be sudden or gradual and the progression is in a stepwise manner. BD can often coexist with Alzheimer's disease. Behaviours that slow the progression or control risk factors of atherosclerosis - high blood pressure, diabetes, - such as eating a healthy diet and keeping healthy wake/sleep schedules, exercising, and avoiding smoking, and limiting alcohol - can also slow the progression of BD.

Dementia associated with corticobasal degeneration

Corticobasal degeneration (CBD), sometimes referred to as corticobasal ganglionic degeneration, is considered to be a part of the spectrum of Frontotemporal dementia (FTD). It is characterised by nerve cell death and atrophy or shrinkage of multiple areas of the brain, including the cerebral cortex and basal ganglia. CBD typically occurs in individuals between the ages of 45-70. Rarely, there is a family history of dementia, psychiatric problems or a movement disorder.

Signs and symptoms

Individuals with CBD usually present with either a movement disorder or cognitive deficits. As the disease progresses, they can go on to develop both types of symptoms.

The movement symptoms can be very similar to Parkinson's disease, with slowness, and rigidity, but tremor is less common. These symptoms do not respond to Parkinson's disease medications.

Many individuals with CBD experience a subtle change in sensation or an inability to make the affected limb follow commands. They may have difficulties completing some specific tasks such as brushing teeth, opening door or using tools such as a can opener.

When it affects the legs, a person can have difficulty dancing, or may show an increased tendency to trip and fall. Other symptoms include involuntary stiffening, twisting or contraction (dystonia), or uncontrolled movement of the affected limb (myoclonus).



The cognitive symptoms associated with CBD include language problems such as word finding problem or naming problem. Reading, writing, and simple mathematical calculations may be impaired.

Personality changes, inappropriate behaviour, or repetitive or compulsive activities are seen in FTD and are common. Short-term memory problems are less common.

Diagnosis

Patients with CBD who present with cognitive symptoms are often initially diagnosed with FTD or Alzheimer's disease. It is when movement symptoms develop that the possibility of CBD is considered. Occasionally, a diagnosis of CBD is not apparent until at autopsy, when a microscopic examination of the brain shows ballooned neurons, and protein inclusions from accumulation of tau protein.

Progression: CBD usually progresses slowly over 6-8 years. Movement symptoms tend to progress slowly from one side of the body to the other or from leg to arm on the same side of the body.

Treatment

There is no specific course of treatment for CBD at this time. Treatment is basically symptomatic. Patients with rigidity and walking difficulty may respond to medications used for treating Parkinson's disease.

Dystonia and myoclonus may respond to muscle relaxants or anti-seizure medications. Memory and behaviour problems may or may not respond to Aricept, a medication for Alzheimer's disease.

Associated depression and or anxiety may respond to antidepressants such as Sertraline, Citalopram etc.

Physical therapy and stretching exercises may be recommended to relieve rigidity, prevent contractions and deformities, and maintain muscle strength.

Assistive devices such as walkers, cane, and speech, physical, and occupational therapy are other helpful strategies to manage movement disorders and language difficulties.



Posterior cortical atrophy (Benson's Disease)

Posterior cortical atrophy (PCA), also called Benson's syndrome, is a form of dementia which is usually considered an atypical variant of Alzheimer's disease. The disease causes atrophy of the posterior part of the cerebral cortex, resulting in the progressive disruption of complex visual processing. The famous author Terry Pratchett suffered with this type of dementia.

The progressive degenerative condition involves the loss and dysfunction of brain cells, and is thought to be behind 5% of cases of Alzheimer's representing thousands of people in the UK.

Although both Alzheimer's and PCA involve the loss and dysfunction of brain cells, they affect different parts of the brain. Typically Alzheimer's disease first impacts the sides of the brain – areas which play an important role in memory, which is why memory problems are usually the first symptom.

In PCA however, the disease first affects the back of the brain, known as the occipital lobe. This part of the brain is responsible for vision, so people with PCA often initially experienced visual problems.

Rather than being based on memory where people forget appointments and where they've put things, for people with PCA the first thing they notice is complex visual behaviour.

"They might find they're clipping wing mirrors when they're driving or having problems parallel parking. When they're reading, words will jump around and a lot of people will have difficulty perceiving objects such as glass doors."

The first symptoms of PCA tend to occur when people are in their late 40s, 50s or early 60s, so it has an earlier onset than Alzheimer's disease. But the first signs are often subtle and so it may be some time before a formal diagnosis is made.

As damage in the brain spreads and the disease progresses, people develop the typical symptoms of Alzheimer's disease such as memory loss and confusion.

There is no specific treatment for PCA but some people use medications used for Alzheimer's disease. And like Alzheimer's, there's no cure.

PCA is said to be the most poorly understood type of dementia. This is because the changes in the brain are very similar to those that happen in Alzheimer's.



The same changes in brain cells happen, the same proteins build up, and they have the same pathology, but we simply don't know why it affects different parts of the brain."

Sir Terry himself described PCA as giving him the "opposite of a superpower" in a comment piece he wrote on assisted dying for the Guardian in February 2010.

"Sometimes I cannot see what is there," he wrote.

"I see the teacup with my eyes, but my brain refuses to send me the teacup message. It's very Zen. First, there is no teacup and then, because I know there is a teacup, the teacup will appear the next time I look.

"I have little work-arounds to deal with this sort of thing – people with PCA live in a world of work-arounds.

PCA manifests itself through sight problems and difficulty with topological tasks, such as buttoning up a shirt. Typically residents will struggle with interpreting their environment. Trouble with reading, become startled at fast moving objects and have problems judging distances PCA develops slowly.

Time Out

Consider how you would support a residents suffering with PCA to maintain their independence as much as possible?

What factors would you need to consider?



Dementia associated with HIV

HIV (Human Immunodeficiency Virus) is an infection that weakens the immune system, decreasing the ability of the body to fight infections and diseases. HIV infection can affect the brain in up to half of people with HIV. The effects on the brain result in mild cognitive complaints and dementia. Cognitive impairment is common in HIV but dementia is much rarer. Before the use of antiretroviral drugs, around 20-30% of people with advanced HIV infection developed dementia. This has now decreased to around 2%.

Neurocognitive impairment in people with HIV may be caused by the virus directly damaging the brain or could be the result of a weakened immune system enabling infections and cancers to attack the brain.

Symptoms may include problems with short-term memory, language and thinking, difficulties with concentration and decision making, unsteadiness, mood changes and hallucinations. People may also have problems with their sense of smell. Some people may experience only mild cognitive impairment such as a decline in the ability to think quickly or clearly.

HIV is easily overlooked as a possible cause of dementia and, even when someone is known to have HIV infection, cognitive impairment can sometimes be difficult to diagnose because the symptoms are similar to those of other conditions such as depression.

Treatment with a combination of at least three antiretroviral drugs often prevents cognitive impairments worsening and, for many people, can reverse the cognitive damage caused by HIV. Rehabilitation programmes may also help people with HIV-related cognitive impairment to re-learn skills.

Korsakoff syndrome

Korsakoff syndrome is a chronic memory disorder caused by severe deficiency of thiamine (vitamin B-1). Thiamine helps brain cells produce energy from sugar. When levels fall too low, brain cells cannot generate enough energy to function properly.

Korsakoff syndrome is most commonly caused by alcohol misuse, but can also be associated with AIDS, cancers that have spread throughout the body, chronic infections, poor nutrition and certain other conditions.



Korsakoff syndrome is often — but not always — preceded by an episode of Wernicke Encephalopathy, which is an acute brain reaction to severe lack of thiamine. Wernicke Encephalopathy is a medical emergency that causes life-threatening brain disruption, profound confusion, staggering and stumbling, lack of coordination, and abnormal involuntary eye movements.

Because the chronic memory loss of Korsakoff syndrome often follows an episode of Wernicke encephalopathy, the chronic disorder is sometimes known as Wernicke-Korsakoff Syndrome. But Korsakoff syndrome can also develop in individuals who have not had a clear-cut prior episode of Wernicke encephalopathy.

Korsakoff syndrome and its associated thiamine deficiency is not the only mechanism through which heavy drinking may contribute to chronic thinking changes and cognitive decline.

Alcohol misuse may also lead to brain damage through the direct toxic effects of alcohol on brain cells; the biological stress of repeated intoxication and withdrawal; alcohol-related cerebrovascular disease; and head injuries from falls sustained when inebriated.

Prevalence

Scientists don't know exactly how many people have Korsakoff syndrome. It's widely considered less common than Alzheimer's disease, vascular dementia, frontotemporal dementia or dementia with Lewy bodies. Like more common types of dementia, it may be under diagnosed.

Symptoms

Korsakoff syndrome causes problems learning new information, inability to remember recent events and long-term memory gaps. Memory difficulties may be strikingly severe while other thinking and social skills are relatively unaffected. For example, individuals may seem able to carry on a coherent conversation, but moments later be unable to recall that the conversation took place or to whom they spoke.

Those with Korsakoff syndrome may “confabulate,” or make up, information they can't remember. They are not “lying” but may actually believe their invented explanations.



Scientists don't yet understand the mechanism by which Korsakoff syndrome may cause confabulation.

Diagnosis

Korsakoff syndrome is a clinical diagnosis representing a physician's best judgment about the cause of a person's symptoms. There are no specific laboratory tests or neuroimaging procedures to confirm that a person has this disorder. The syndrome may sometimes be hard to identify because it may be masked by symptoms of other conditions common among those who misuse alcohol, including intoxication or withdrawal, infection or head injury.

Experts recommend that a medical workup for memory loss or other cognitive changes always include questions about an individual's alcohol use.

Anyone admitted to the hospital for an alcohol-related condition should be professionally screened for memory loss and cognitive change. The screening should include supplementary questions to assess recent memory.

If screening suggests impairment, the person should receive a more detailed cognitive workup.

Causes and risk factors

Scientists don't yet know exactly how Korsakoff syndrome damages the brain. Research has shown that severe thiamine deficiency disrupts several biochemicals that play key roles in carrying signals among brain cells and in storing and retrieving memories.

These disruptions destroy brain cells and cause widespread microscopic bleeding and scar tissue. Most cases of Korsakoff syndrome result from alcohol misuse.

Scientists don't yet know why heavy drinking causes severe thiamine deficiency in some alcoholics, while others may be affected primarily by alcohol's effects on the liver, stomach, heart, intestines or other body systems.

Researchers have identified several genetic variations that may increase susceptibility to Korsakoff syndrome. Poor nutrition may also raise risk. Korsakoff syndrome may sometimes be associated with disorders other than alcohol misuse, including anorexia, overly-stringent dieting, fasting, starvation or weight-loss surgery; uncontrolled vomiting; AIDS; kidney dialysis; chronic infection; or cancer that has spread throughout the body.



Outcomes

Wernicke encephalopathy, a related disorder that sometimes precedes Korsakoff syndrome, is a medical emergency. Untreated, it causes death in up to 20 percent of cases and progresses to Korsakoff syndrome in 85 percent of survivors. Abnormal eye movements that occur in Wernicke encephalopathy may respond to injectable thiamine within a few days.

Lack of coordination and clumsiness may begin to improve after about a week but may take several months to clear up completely. Confusion also takes several months to clear up. As confusion clears, the severe memory problems associated with Korsakoff syndrome may become more noticeable.

In those who develop Korsakoff syndrome, with or without a preceding episode of Wernicke encephalopathy, there are few studies on long-term outcomes. Available data suggest that about 25 percent of those who develop Korsakoff syndrome eventually recover, about half improve but don't recover completely, and about 25 percent remain unchanged.

Some research suggests that those who recover from an episode may have a normal life expectancy if they abstain from alcohol.

Treatment

Some experts recommend that heavy drinkers and others at risk of thiamine deficiency take oral supplements of thiamine and other vitamins under their doctor's supervision.

Many experts also recommend that anyone with a history of heavy alcohol use who experiences symptoms associated with Wernicke encephalopathy, including acute confusion, prolonged nausea and vomiting, unusual fatigue or weakness, or low body temperature or blood pressure, be given injectable thiamine until the clinical picture grows clearer.

Once acute symptoms improve, individuals should be carefully evaluated to determine if their medical history, alcohol use and pattern of memory problems may be consistent with Korsakoff syndrome.

For those who develop Korsakoff syndrome, extended treatment with oral thiamine, other vitamins and magnesium may increase chances of symptom improvement.



Abstaining from alcohol is a cornerstone of effective long-term treatment. Those with Korsakoff syndrome have a reduced tolerance for alcohol and may be at high risk for further alcohol-related health problems.

What is Huntington's disease and how is it associated with dementia?

Huntington's disease is a progressive brain disorder caused by a gene on chromosome 4. It is an "autosomal dominant disease," meaning if a person has inherited the gene from a parent with Huntington's disease, they will eventually develop the disease as well. It creates a progressive decline in thinking and memory, uncontrolled movements of the body and changes in mood and behaviour.

What are the symptoms?

Symptoms of Huntington's disease typically appear between 30-50 years of age, but can be present as early as age 2 or as late as 80 years. Common symptoms include uncontrolled movement of the arms, legs, head, face and upper body; and a significant decline in thinking and reasoning including memory, concentration, judgment and the ability to plan and organize. The brain changes in Huntington's disease also lead to alterations in mood, especially depression, anxiety and unusual anger and irritability. Another common symptom is obsessive-compulsive behaviour, where a person repeats an action or a question over and over again.

How is it diagnosed?

The gene that causes Huntington's disease was identified in 1993. Genetic testing is now available to confirm the presence of the gene in symptomatic people with suspected symptoms and those without symptoms, but who are determined to be at risk of developing it based on a family history of the disease.

What kind of treatment is available?

As with Alzheimer's disease, there is currently no cure for Huntington's disease and no way to slow or stop the progressive changes in the brain. Treatments focus on pharmaceutical drugs to help manage symptoms (involuntary movements or "chorea," irritability and obsessive-compulsive thoughts/actions).



Other symptoms such as anxiety, depression and insomnia, should be treated according to generally accepted guidelines. Experts recommend people with Huntington's to keep their medical appointments and not to become discouraged if their health care team requires additional time to find the best pharmaceuticals and the most effective doses for the person.

What is Down syndrome dementia?

Down syndrome (Trisomy 21) is a condition characterized by the presence of extra material on chromosome 21. People living with Down syndrome have an increased risk of developing dementia as they get older. Dementia associated with Down syndrome is thought to be very similar to traditional forms of Alzheimer's disease.

More than 75% of people with Down syndrome aged 65 and older are living with Alzheimer's disease, about 6 times the number of people in the greater population in the same age group.

The risk of developing Alzheimer's disease in individuals with Down syndrome has been found to increase with advancing age.

Since people with Down syndrome typically live up to 55-65 years of age, they are more likely to have young-onset Alzheimer's disease rather than the classical type that affects individuals older than 65 years.

People with Down syndrome have been found to have significant levels of plaques and tangles in brain nerve cells – a common feature of Alzheimer's disease – appearing by the age of 40. Surprisingly enough, not everyone with these brain changes develop symptoms of Alzheimer's disease. It is not known why some people with Down syndrome who have brain changes develop symptoms - while others do not.

What are the symptoms?

In people living with Down syndrome, changes in overall function, personality and behaviour may be more common as early signs of Alzheimer's disease than memory loss. Early symptoms may include:

- lack of interest in socializing, communicating or expressing thoughts
- lack of initiative and enthusiasm for common activities



- decreased ability to focus or concentrate
- emotional changes such as sadness, fearfulness, anxiety or irritability
- behavioural changes including aggression, restlessness or sleep disturbances
- seizures that begin in adulthood
- changes in coordination and walking
- increased noisiness or excitability.

Researchers are unclear as to why these symptoms differ for people with Down syndrome compared to the general population.

Genetic component ? Can it be passed on to family members?

It is believed that the extra chromosome 21 is responsible for the symptoms of dementia in individuals with Down syndrome. One of the chromosome 21 genes in Down syndrome codes for amyloid precursor protein (APP). The exact function of APP is not known, but it is believed that day-to-day brain activity involves continuous "processing" of APP into shorter pieces. One of the brain's APP processing pathways produces beta-amyloid, the chief component of plaques and a prime suspect in Alzheimer's disease. Having an extra copy of the APP gene may increase the production of beta-amyloid and trigger the chain of biological events leading to Alzheimer's disease.

How is it diagnosed?

Diagnosing dementia in a person with Down syndrome can be difficult because of the challenges involved in assessing changes in cognitive skills in people with intellectual disabilities. The following guidelines have been recommended:

- **document baseline adult function by age 35.** Ideally, each individual's medical record should include detailed information about adult abilities and intellectual, social and behavioural functions
- **watch for changes in day-to-day function.** Reduced enthusiasm for daily activities, lack of interest in social interactions and changes in personality and behaviour are often early signs
- **consider professional assessment by a dementia expert.** A variety of cognitive tests have been used to evaluate thinking changes in adults with Down syndrome. Keep in mind that cognitive tests should never be used as the only benchmark to diagnose dementia in a person with Down syndrome



- **rule out other causes of symptoms.** Thyroid conditions, depression, chronic ear and sinus infections and sleep apnea can cause changes in a person's thinking and functioning.

What kind of treatment is available?

Currently there are no approved pharmaceutical drugs used specifically to treat dementia associated with Down syndrome. In the United Kingdom, cholinesterase inhibitors (a class of medications approved to treat Alzheimer's disease), are approved to treat dementia in people with Down syndrome. However, there isn't enough evidence to reach a definitive conclusion about their effectiveness.

An international clinical study has shown no benefit for the Alzheimer's drug Memantine (Ebixa) in adults with Down syndrome. More research and clinical studies are required to identify effective treatments for dementia in those with Down syndrome.

What is Parkinson's disease dementia?

Parkinson's disease dementia (PDD) refers to a decline in thinking and reasoning that eventually affects a number of people with Parkinson's disease. Parkinson's disease is a fairly common neurological disorder, estimated to affect nearly 2% of people older than age 65 years of age. It is estimated that 50 – 80% of those with Parkinson's disease eventually experience Parkinson's disease dementia. The average time from onset of Parkinson's disease to developing dementia is about 10 years.

The brain changes linked to Parkinson's disease and Parkinson's disease dementia (PDD) are irregular, tiny deposits of alpha-synuclein protein called Lewy bodies.

Lewy bodies are also found in several other brain disorders, including Lewy body dementia. Evidence suggests that Lewy Body dementia, Parkinson's disease and PDD may be linked to the same underlying changes in brain processing.

In fact, many experts believe that Parkinson's Disease Dementia and Lewy body dementia are two different outcomes of the same underlying problems with brain processing - but most experts recommend continuing to diagnose them as separate disorders.



Another puzzling factor is that many people with these two types of dementia also have the common features of Alzheimer's disease – plaques and tangles in brain nerve cells.

What changes can be expected / common symptoms

- changes in memory, concentration and judgement;
- problems with visual perception; muffled speech;
- visual hallucinations;
- delusions or paranoia;
- depression; irritability and anxiety,
- sleep disturbances, including excessive daytime drowsiness
- rapid eye movement (REM) sleep disorder.

Like other types of dementia, PDD may get worse over time and the speed of progression can vary from individual to individual.

What are the risk factors?

Certain factors at the time of Parkinson's disease diagnosis may increase a person's future risk of developing dementia - including older age, greater severity of motor symptoms and having mild cognitive impairment. Additional risk factors may include: hallucinations in a person who doesn't yet have other dementia symptoms; excessive daytime sleepiness; postural instability and gait disturbance (PIGD) which includes 'freezing' in mid-step, difficulty initiating movement, shuffling, problems with balance and falling.

How is it diagnosed?

Unfortunately, there is currently no single test, or combination of tests, that conclusively determines the presence of Parkinson's disease dementia (PDD). Guidelines for diagnosing PDD and Lewy body dementia are:

- **A diagnosis of PDD** is when a person is originally diagnosed with Parkinson's based on movement symptoms and dementia symptoms don't appear until one year (or more) has passed.



- **Diagnosis of Lewy body dementia** is made when 1) Dementia symptoms consistent with Lewy body dementia develop first; 2) Both dementia symptoms and movement symptoms are present at the time of diagnosis; and 3) When movement symptoms develop within a year of Lewy body dementia diagnosis.

Since individuals with Parkinson's disease are at a higher risk for dementia as their disease progresses, doctors monitor those with Parkinson's closely for signs of changes. When someone with Parkinson's disease develops changes in thinking, doctors often request a magnetic resonance imaging (MRI) scan of the brain to rule out tumors, structural changes and evidence of vascular disease.

What kind of treatment is available?

There are currently no treatments to slow or stop the brain cell damage caused by Parkinson's disease dementia (PDD). Current treatment strategies focus on alleviating or managing the symptoms.

Treatment considerations involving medications might include the following:

- Cholinesterase Inhibitors, a common approach to treating Alzheimer's disease and could also assist with PDD symptoms. Antipsychotic drugs should be used with extreme caution in PDD, since they may cause serious side effects in as many as 50% of those with PDD and Lewy body dementia. Side effects may include sudden changes in consciousness, impaired swallowing, acute confusion, episodes of delusions or hallucinations, or the appearance of worsening Parkinson's symptoms.
- L-dopa may be prescribed to treat Parkinson's movement symptoms, but is thought to aggravate hallucinations and confusion in those with PDD and Lewy body dementia.
- Antidepressants may be used to treat depression, which is common in both PDD and Lewy body dementia.
- Clonazepam may be prescribed to treat REM sleep disorder.



Dementia associated with Multiple Sclerosis

Some people with multiple sclerosis (MS) experience some change in their mental abilities depending on the part of the brain affected by the disease. Individuals may be affected in different ways and to different degrees, over a period of time. The mental abilities most likely to be affected are as follows:

- memory
- concentration
- ability to problem solve
- emotional problems
- mood swings.

The decline in mental abilities associated with MS is not usually severe enough to be categorized as dementia. It is more typical to describe the symptoms as cognitive difficulties. For more information, please contact the MS Society.



Acknowledgements

Source: www.alzheimers.org.uk

National Institute of Neurological Disorders and Stroke:

http://www.ninds.nih.gov/disorders/corticobasal_degeneration/corticobasal_degeneration.htm



