Module 2:
Diagnosing frontotemporal dementia

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Frontotemporal dementia (FTD) poses a diagnostic challenge for health professions. Affected individuals may initially present to their general practitioners or local medical officers when symptoms emerge. Others may be seen in specialist clinics relatively early in the course of the disease. The condition, however, is often not recognised and is frequently misdiagnosed. Thus, the road to diagnosis has traditionally been long and difficult.

Many carers of people with FTD indicate that they have difficulty convincing health professionals to investigate the symptoms they are reporting. This experience may be explained by the following:

- Many health professionals may not have been exposed to people with younger onset dementias, and therefore do not consider dementia as a differential diagnosis for those younger than 70 years of age.

- Early changes in FTD are often subtle and not detected by routine cognitive screening tools. Such tools tend to be insensitive to the executive function and language changes commonly seen in FTD.

- Many of the early emotional, behavioural, and relationship changes seen in FTD may appear to have a psychiatric or social cause. In younger people, these are more common than FTD and so referrals for psychotherapy or counselling may be the first course of action.

- Given FTD is a progressive neurological condition with limited treatment options to date, many Health Professionals may be reluctant to label someone with FTD until they have exhausted all other possibilities. Whilst this is appropriate to some degree, this wait and watch approach may take longer than many carers are willing to wait for answers. Hence, it is important to consider early specialists referral to minimise delays for carers in getting the answers they seek.

It is hoped that, in light of the information provided in this toolkit, more health professionals will consider the differential diagnosis of FTD when people under 70 years of age present with the symptoms described in *Module 1 – What is Frontotemporal dementia*? The goal of the present module is to provide health professionals with information designed to assist in identifying possible cases of FTD and referring suspected cases for further investigation.

Early diagnosis of progressive conditions such as FTD is critical. In 2003, Hodges and colleagues [1] reported an average delay of three years from symptom onset to FTD

diagnosis across two specialist centres. During the period when the illness remains undiagnosed, people with these symptoms and their carers are likely to experience distress and confusion. It is hoped that raising awareness of the signs of FTD will also reduce the delay to diagnosis, thereby enabling earlier intervention.

Given that there is no cure for FTD at this time, education and support are currently recognised as the most important interventions we can provide for patients and carers of people with FTD. Early access to knowledge and support should lead to improved quality of life and outcomes for patients and caregivers. If early diagnosis is achieved, it is also possible that the person with FTD may retain some capacity to actively participate in organising his or her affairs and planning for future care needs.

Meeting the diagnostic challenge posed by FTD requires the collection and integration of specific information from a number of sources. Information from a single source such as a cognitive assessment or neuroimaging is insufficient to accurately identify FTD. Thus, diagnosis is ideally informed by a multidisciplinary team. The information required for diagnosis, the contributions made by the relevant specialities, and specific contact details for specialist centres in Australia are described here. Important post-diagnostic considerations are also outlined.

Specialist inputs

Input from the following healthcare specialties can contribute to a diagnosis of FTD:

- Neurology
- Neuropsychiatry
- Clinical Neuropsychology
- Psychiatry
- Geriatric Medicine
- Speech Pathologist, Occupational Therapist, Cognition Nurse or Social worker,
- Radiology
- Neuropathology
- Genetics

A medical specialist in dementia diagnosis such as a neurologist, neuropsychiatrist or geriatrician will often oversee the process of gathering the relevant diagnostic information from the other specialties. He or she will take all sources of information into account, looking for patterns supportive of a diagnosis of FTD after ruling out other potentially treatable causes of the person's symptoms. This information should also be fed back to the person's general practitioner. The diagnosis of FTD, however, is ideally made by a specialist who has experience in this area. The information critical to the diagnostic formulation is described below.

Essential diagnostic information

Collaborative assessment and symptom history

When a person presents with symptoms suggestive of FTD, the most important diagnostic information is a clear history of the nature, onset, and progression of symptoms. All cases of FTD are characterised by an insidious onset and gradual progression, despite variability in the rate of symptom progression. The onset of symptoms may be difficult to pinpoint [2], but a sudden onset of symptoms is not consistent with a diagnosis of FTD.

The importance of independent interviews

It is important to take a careful history from the person. It is essential, however, to speak at length with knowledgeable informants. People with FTD may present with early changes in insight and may inadvertently provide misleading information [2]. Those providing a collaborative history, such as family members should not just be seen in the presence of the person. The nature of the symptoms (e.g., embarrassing or disturbing) may be such that family and friends do not feel comfortable providing honest descriptions in front of their loved ones. It is essential, therefore, that interviews with informants take place independently.

Complaints supportive of a diagnosis of FTD include uncharacteristic behaviour, inappropriate social conduct, loss of judgement, changes in activity levels, language difficulties, and subtle or obvious personality changes (see Module 1 – *Clinical Symptoms* for a comprehensive list of possibilities). It is important that complaints such as changes in the person's sexual behaviour, empathy, or motivation are queried in detail. It is not uncommon for such symptoms to be interpreted as signs of relationship discord and referrals made to marriage counselling, when these symptoms may be the early signs of a neurodegenerative disorder.

The assessing clinician should also take care to understand the premorbid personality characteristics, tendencies, and abilities of the patient. Many symptoms of FTD reflect exaggeration of premorbid idiosyncratic traits and may only be conceptualised as such when the previously well person is understood.

Finally, a clear family history of dementia and other neurological conditions should be obtained. While family histories are not diagnostic in and of themselves, a positive history can be telling in combination with other supportive features. Even when there is no family history of FTD, clinicians should be aware that FTD was not often diagnosed in previous generations [2] or was diagnosed as another disorder such as Alzheimer's disease.

Structured assessment tools may be particularly helpful in identifying behavioural and other functional changes (see Measures and screening tools).

Physical examination

A medical specialist may perform a thorough physical and neurological examination, order blood tests, and/or carry out further investigations of the body in order to rule out potentially treatable causes of functional decline. Given the possible presence of MND or other movement disorders in FTD, it is particularly important to check for the presence of motor symptoms during these examinations. A swallowing assessment may be recommended in order to ensure that nutritional needs are met while the danger of aspirating food or drink is avoided.

Cognitive assessment

A thorough cognitive assessment is a critical component of the diagnostic process in suspected FTD. This is usually performed by an experienced clinical neuropsychologist. Neuropsychologists are psychologists who have postgraduate training in the specialist area of neuropsychology. They have expertise in brain-behaviour relationships and are able to measure the impact of various disease processes on cognition and behaviour. The role of the neuropsychologist in the diagnosis of FTD is to characterise the patient's cognitive abilities and to identify changes in cognition. The neuropsychologist establishes a history and pattern of cognitive, behavioural, and emotional changes, which can critically inform the team's diagnostic reasoning.

The most important cognitive domains to assess in suspected FTD are executive functions and language. Executive functions are those skills required for purposeful, planned, goal-directed activity. They are also the skills which promote adaptive function in everyday life. These cognitive processes are mediated by the frontal lobes and impairments in these skills are a hallmark of FTD. A speech pathologist may add significant expertise in characterising the nature of speech production, communication and language changes.

Given that some forms of FTD involve early changes in particular aspects of language, the neuropsychologist must also carefully investigate any potential changes in language skills. These changes may be quite subtle in the early stages of the disease and require careful examination if they are to be properly identified.

Importantly, many individuals can perform well on a variety of formal cognitive tests even in the presence of significant behavioural symptoms [2-4]. This is particularly true within the domain of executive function. Some executive dysfunction only manifests in behavioural ways within the natural environment rather than on structured cognitive testing. Thus, a holistic approach to assessment is essential.

Additional domains that a neuropsychologist may elect to investigate include activities of daily living (which may be particularly impaired in FTD [2]), everyday behavioural function, and social cognition.

The insensitivity of routine cognitive screening

Primary care physicians (GP's) may also be interested in conducting cognitive screening before referring patients to specialists. A mismatch between the nature and degree of impairments in FTD and the demands of various screening tools may, however, result in misleading results [5]. It is important to note that simple cognitive screening tools such as the Mini-Mental State Examination (MMSE) are not sufficient to assess the status of individuals suspected of having FTD [6, 7]. People with FTD can even obtain a perfect score on the MMSE, as this measure can be insensitive to executive dysfunction.

It is critical for all practitioners to use tools specifically designed to assess cognition in FTD, or tools which are known to be sensitive to the changes typical of FTD (see Measures and screening tools).

Neuroimaging

Neuroimaging is increasingly utilised in the diagnosis of FTD. Brain scans alone are not sufficient to diagnose the condition, but high quality imaging can contribute important information to the diagnostic formulation. Neuroimaging is also useful in ruling out differential diagnoses such as intracranial tumours (see *Differential Diagnoses*).

Different imaging modalities may be used to examine different aspects of the brain's integrity, and can help detect structural pathology, white matter changes, atrophy, and functional changes such as reduced blood flow or hypometabolism [8]. Structural modalities include computed tomography (CT) and volumetric magnetic resonance imaging (MRI). CT scans are readily available and relatively inexpensive; they do not, however, offer as much precision as MRI scans. CT scans are useful in detecting changes such as strokes and tumours, but MRI scans are better suited to detecting the more subtle changes that often accompany insidious dementia processes such as FTD. Importantly, even structural MRI scans are not sufficiently sensitive to detect some of the earliest changes, which may only be manifested at a functional level [8].

Functional imaging techniques include single photon emission computed tomography (SPECT), positron emission tomography (PET), and functional MRI (fMRI). These scans are useful in detecting hypometabolism or hypoperfusion, respectively, in regions of interest. Even if structural brain changes are not evident on structural MRI, these scans may reveal reduced functionality of brain tissue. In the case of FTD, this reduced function may eventually translate into atrophy that can be visualised on structural imaging.

Functional imaging is generally more expensive and less available than structural scanning. Thus, it may only be offered in tertiary referral centres or in the context of research [8].

In most cases of FTD, focal abnormalities of the frontal and anterior temporal lobes are evident on neuroimaging. This may be symmetrical or asymmetrical. Patients with bvFTD often have both frontal and temporal lobe abnormalities [9], with particular involvement of the ventromedial frontal region [9]. In the case of language symptoms, there is a direct link between symptom type and site of anatomic brain damage [10]. PNFA tends to be associated with atrophy or dysfunction of the language-dominant (usually left) inferior frontal lobe and anterior insula [10, 11]. In SD, bilateral abnormalities in the inferior and lateral anterior temporal regions are typically evident, and are usually worse on the left [10]. Atrophy of the basal ganglia and loss of pigmentation in the substantia nigra of the brain stem may also be evident in some cases of FTD [2, 12], and may relate to movement symptoms. A visual rating scale has recently been developed which may assist in interpreting structural imaging in suspected FTD [3].

It is important to emphasise that the brains of some individuals with FTD do not show clear atrophy beyond that expected for their age. Early abnormalities may only be detected on functional neuroimaging, or may not be detectable with currently available technology until later in the course of the disease [13].

Neuropathological testing

Tests of cerebrospinal fluid and/or plasma protein levels may be used by some clinicians to help identify FTD, though diagnostically specific biomarkers have yet to be identified [11]. At most, such testing may assist with distinguishing FTD from dementia of the Alzheimer's type (DAT) [2, 11].

Brain banking – a final answer

Biopsies of brain tissue are rarely obtained during the patient's life due to the invasive nature and limited benefit of such procedures. The most important contribution of neuropathological testing in suspected FTD occurs at post-mortem (post death,) when characterisation of the brain and the neuropathological changes present in samples of brain tissue can be performed [12]. Such an examination provides a definitive diagnosis of the biological disease process underlying clinical FTD.

Brain bank availability varies from state to state in Australia. Details of some brain banks are listed below.

Victoria & Tasmania
Victorian Brain Bank Network
Mental Health Research Institute of Victoria
Director: Professor Catriona McLean
Coordinator: Ms. Fairlie Hinton
Phone: 03 8344 1900
Email: f.hinton@mhri.edu.au
Website: www.mhri.edu.au/brain-bank

New South Wales & Australian Capital Territory
Sydney Brain Bank
Neuroscience Research Australia (Prince of Wales Medical Research Institute)
Sydney, NSW
Director: Glenda Halliday
Liaison Officer: 02 9399 1107
Email: braindonors@neura.edu.au
Website: http://www.neura.edu.au/sydneybrainbank

Queensland
Queensland Brain Bank
School of Chemistry & Molecular Biosciences, The University of Queensland
Brisbane, QLD
Coordinator: Ms. Alison Eckert
Phone: 07 3365 4614
Email: a.eckert@uq.edu.au

South Australia & Northern Territory
South Australian Brain Bank
Coordinator: Ms. Robyn Flook
Phone: 08 8204 4107
Email: Robyn.Flook@flinders.edu.au

Western Australia
Royal Perth Hospital
Coordinator: Ms. Caroline Caseley
Phone: 08 9224 1192
Email: caroline.casely@health.wa.gov.au

Further enquiries:
Australian Brain Bank Network
National Neuroscience Facility
Level 3, 161 Barry Street
Carlton South, Victoria 3053
Director: Professor Catriona McLean
Coordinator: Ms. Fairlie Hinton
Phone: 03 8344 1900
Fax: 03 9349 5105
Email: f.hinton@mhri.edu.au
Website: http://www.nnf.com.au/platforms/abbn/

Genetic screening

The FRONTIER research group recommends that people with a first degree relative who suffered from a disease on the FTD spectrum be screened for *MAPT* and *GRN* mutations after genetic counselling [2]. Other researchers agree that the benefit of genetic screening depends on the strength of family history and on the clinical variant [11].

Genetic testing is problematic in many parts of Australia as it is not covered by Medicare and may only be available at certain centres. It is important to discuss this option with the treating specialist. Any family members seeking testing should be referred to an expert neuro-genetics counsellor for advice.

Measures and screening tools

A number of useful screening tools have been developed to assist clinicians in diagnosing FTD. The list below is organised according to the functional domain targeted by each measure, and by alphabetical order within categories.

Cognitive screening

Addenbrooke's Cognitive Examination (ACE) [14] *and Addenbrooke's Cognitive Examination Revised (ACE-R)* [15] are bedside test batteries that assess a number of cognitive domains and incorporate the *MMSE* [16]. Both have been shown to be effective in differentiating FTD from Alzheimer's Disease (AD) in mildly demented patients [14, 15]. Administration time: 12-20 minutes. Test materials, administration details, and normative data are available for free download from the FRONTIER research group website: http://www.ftdrg.org/research/test-downloads/

The Frontal Assessment Battery (FAB) [17] is a brief bedside battery designed to assess frontal lobe functions using tests of cognition and motor behaviour. It has been shown to differentiate between controls and patients with frontal lobe dysfunction (including patients with FTD), though its ability to distinguish between patients with frontal and those with non-frontal lobe injuries has not been tested. Some suggest that FAB performance may not reflect frontal function exclusively [18]. Administration time: 10 minutes. Content, administration instructions, and scoring details published in the journal *Neurology* [17].

INECO Frontal Screening (IFS) [18] is a brief battery designed to assess executive functions which incorporates some items from the FAB as well as additional measures. It has been shown to discriminate between controls and patients with dementia, and between patients with bvFTD and AD [18]. The authors recommend using the IFS in combination with the ACE to perform a more comprehensive cognitive screen. Administration time: 10 minutes. Content, administration instructions, and scoring details published in the *Journal of the International Neuropsychological Society* [18].

Additional domain-specific cognitive tests which may be of interest to clinical neuropsychologists are available for free download from the FRONTIER research group website: http://www.ftdrg.org/research/test-downloads

Behaviour screening

The *Cambridge Behavioural Inventory (CBI)* [19, 20] is a carer questionnaire designed specifically to capture the behavioural and cognitive changes characteristic of both FTD and AD, and has been shown to discriminate between the two [19, 21]. It incorporates some aspects of the generic *Neuropsychiatric Inventory* (see below) as well as additional questions related to common symptoms in FTD and to activities of daily living. The original 80-item questionnaire and a 45-item revised version (CBI-R) are both available for free download from the FRONTIER research group website:
http://www.ftdrg.org/research/test-downloads
Normative data may be accessed in supporting publications [19, 20, 22].

The Behavioural Frontotemporal Lobe Dysfunction Assessment Scale [23] is a structured interview-based rating scale designed to be administered to caregivers, and has been shown to distinguish between FTD, AD, and vascular dementia [23]. Content and interpretation guidelines are published in the journal *Alzheimer's Disease and Associated Disorders* [23].

The Frontal Behavioural Inventory (FBI) [24] is a 24-item caregiver questionnaire specifically developed to assess the behavioural disturbances of FTD. It has been shown to discriminate between different FTD phenotypes and between FTD and other forms of dementia [24-26]. Content, scoring instructions, and interpretation guidelines are published in the *Canadian Journal of Neurological Sciences* [24].

The Neuropsychiatric Inventory (NPI) [27] is a caregiver-based structured interview designed to briefly assess problematic behaviours and psychopathology in dementia. It has been shown to be effective in discriminating many cases of FTD from AD and vascular dementia [28], but may not be as effective as the FBI in this regard [25]. Interview content and scoring instructions are described in the journal *Neurology* [27], and the full instrument will be provided by the authors upon request.

The Stereotypy Rating Inventory (SRI) [29] is a brief structured interview designed to assess the severity and frequency of five stereotypic behavioural disturbances characteristic of FTD. It has been shown to distinguish FTD from AD and vascular dementia, and may also be useful in monitoring the effects of therapies designed to treat stereotypic behaviours.

An *Informant-based Questionnaire* [30] was developed by the Manchester group with the specific aim of distinguishing cases of FTD from AD using retrospective reports, even after the patient's death, in order to clarify family histories of dementia. This may be useful in making judgements about a patient's familial risk and/or whether genetic screening is indicated. It is published in the *Journal of Neurology, Neurosurgery, and Psychiatry* [30].

Severity and decline

The FTDL-specific Clinical Dementia Rating (FTLD-CDR) [31] is a specialised adaptation of the widely used Clinical Dementia Rating (CDR) and provides information specific to the language and behaviour impairments of FTD, as the original CDR tends to underestimate disease severity in FTD [2, 32]. The CDR is published in the journal *Neurology* [33] and details of the Language and Behaviour, Comportment, and Personality domains of the FTLD-CDR are published in the journal *Brain* [31].

The Frontotemporal Dementia Rating Scale (FRS) [32] is a brief, informant-based interview that was recently developed to stage changes in behaviour and activities of daily living in FTD patients. It has been shown to be effective in identifying decline in multiple clinical variants [32]. Administration time: 15 minutes. The FRS is available for free download from the FRONTIER research group website: http://www.ftdrg.org/research/test-downloads/.

Differential diagnoses

A number of differential diagnoses are considered when a patient presents with some of the features of FTD. These conditions might include but are not limited to:

Neurological disorders

- Alzheimer's disease
- Vascular dementia
- Dementia with Lewy Bodies
- Parkinson's disease
- Huntington's disease
- Tumours or tumour-related antibody diseases (paraneoplastic disorders)
- Creutzfeldt-Jakob disease

Psychiatric disorders

- Schizophrenia
- Depression
- Mania
- Anxiety
- Personality disorder
- Asperger's syndrome
- Alcohol and other substance abuse

Many reports have been published regarding the differentiation of FTD from AD Affective blunting, lack of concern, reduced insight, social disinhibition and reduced social awareness, euphoria, stereotypical and aberrant motor behaviour, changes in eating preference and/or behaviours, reduced speech output, and preserved spatial orientation have all been described as effective discriminators of FTD from AD [2, 9, 11, 34].

Referral pathways

The preferred referral pathway for investigation of suspected FTD is a tiered process. The information below is provided in order to facilitate appropriate and timely referrals.

The general practitioner (GP) is usually the first port of call when a medical or mental illness is suspected in a community-dwelling individual. The GP is also likely to be one of the health professionals who will provide ongoing care and contact throughout the course of the disease.

Given the unique and challenging nature of some forms of dementia such as FTD, GPs normally refer patients to specialists for comprehensive assessment when a neurodegenerative process is queried. Referrals may be made to neurologists, geriatricians, or psychiatrists for further investigation.

Within the state of Victoria, GPs are able to refer to the *Cognitive, Dementia, and Memory Service* (CDAMS), a government funded diagnostic service currently unique to Victoria.

First line centres when dementia is queried (Victoria only)

Cognitive, Dementia, and Memory Service (CDAMS)

Ballarat (1)
McKellar Centre
Phone: 03 5279 2344
Fax. 5279 2260

Ballarat (2)
Queen Elizabeth Centre
Phone: 03 5320 3704

Bendigo
John Lindell Rehabilitation Unit
Phone: 03 5454 8500

Bundoora
Bundoora Extended Care Centre
Phone: 03 9495 3272

Burwood
Peter James Centre
Phone: 03 9881 1867

Caulfield (1)
Alfred Health
Ph: 03 9076 6776
Fax: 03 9076 6773
Email: access@cgmc.org.au

Caulfield (2)
Caulfield General Medical Centre
Phone: 03 9276 6010

Cheltenham
Kingston Centre
Phone: 03 9265 1090
Fax: 03 9265 1297
E: SACS_I&A@southernhealth.org.au

Drouin
Baw Baw Health and Community Care Centre
Phone: 03 5625 0242

Heidelberg West
Heidelberg Repatriation Hospital
Phone: 03 9496 2795
Fax: 03 9496 2219

Horsham
Arapiles Building Wimmera Base Hospital
Phone: 03 5381 9333

Hume
Wangaratta, Shepparton, Seymour
Phone: 03 5722 1663
Fax: 03 5721 5605

Kew
St George's Health Service
Phone: 03 9268 0522

Melbourne
Royal Melbourne Hospital
Phone:
Fax: 03 8387 2217
Electronic: www.connectingcare.com

Mt Eliza
Mt Eliza Centre
Phone: 03 9788 1377
Fax: 03 9787 9954

Parkville
Melbourne Extended Care Centre
Phone: 03 8387 2194

St Albans
Sunshine Hospital
Phone: 03 8345 1246

Wantirna
Wantirna Health
Phone: 03 9955 1230
Fax: 03 9955 1388

Australian centres specialising in the diagnosis of FTD

It may be the case that even diagnostic services specialising in dementia do not have extensive experience working with FTD patients, or may find the diagnosis difficult to make. Some examples of centres with particular skills in diagnosing and managing FTD are therefore provided here. This list is not exhaustive. GPs and other health professionals may refer directly to the below centres. Some also accept direct referrals from patients and family members, and some accept referrals for patients in other states and/or countries.

Victoria

Eastern Cognitive Disorders Clinic (ECDC)
Box Hill Hospital
Dr Amy Brodtmann
Eastern Neurosciences
Nelson Road
Box Hill, VIC 3128
Phone: 03 9895 3353 (outpatients)
Fax: 03 9895 4852 (outpatients)
Referral details at: www.ecdc.org.au

Where possible, we ask that the referral be accompanied by a copy of any MRI brain scan reports and results of other investigations, together with details of the clinical history and onset of symptoms.

Neuropsychiatry Unit
Royal Melbourne Hospital
Dr. Denis Velakoulis
John Cade Building,
Level 2, John Cade Building
Cnr Grattan St & Royal Parade
Phone: 03 9342 8750
Fax: 03 9342 8483
Electronic referrals: www.neuropsychiatry.com.au

New South Wales

Frontotemporal Dementia Research Group (FRONTIER)
Professor John Hodges
PO Box 1165
Randwick
Sydney 2031
Phone (personal assistant to Prof Hodges): 02 9399 1134
Fax: 02 9399 1047

Where possible, we ask that the referral be accompanied by a copy of any MRI brain scan reports and results of other investigations, together with details of the clinical history and onset of symptoms.

Referral criteria:
http://www.ftdrg.org/wp-content/uploads/frontier-referral-criteria2.pdf

Western Australia

Dr. Peter Panegyres
Neurodegenerative Disorders Research Pty, Ltd
Perth
Phone: 08 6380 2255
Fax: 08 6380 2055
Email: subiaco@panegyres.com.au
Website: http://www.panegyres.com.au/index.asp

Diagnostic and post-diagnostic considerations

Delivering the diagnosis

People with FTD and their families are greatly assisted if the diagnosing physician is able to provide objective and honest information about FTD and its ramifications at the time of diagnosis. Repetition can also be important due to the often overwhelming nature of the diagnosis [35]. Education regarding the specific nature of this relatively unfamiliar condition can help relieve anxiety about the many unknowns. Information about resources available to assist families in coping with the disease is also critical, including practical details regarding pensions and other financial remuneration options. The information available in this toolkit may serve this purpose, particularly if accompanied by supportive discussion and expert opinion.

Ongoing management

Specialist consultants who diagnose FTD can assist General Practitioners by providing detailed information about the disease. Neurologists, psychiatrists, and geriatricians with experience in FTD can also play a vital role in the ongoing management of the complex symptoms of this condition.

An important aspect of the ongoing management of this disease is the provision of support for carers of people with FTD. It is helpful if the significant burden being accepted by the person's carer is acknowledged and if the carers are also supported and monitored. Carers may experience shock, confusion, grief, and/or depression during the course of the person's illness. Their role in the effective management of the disease is critical and care must be taken to ensure their health and safety.

Carers may benefit from individual counselling, attending support groups, and/or the provision of respite care for their loved ones. The Eastern Cognitive Disorders Clinic in Victoria, FRONTIER in New South Wales, and Alzheimer's Australia – Western Australia run FTD-specific support groups for carers. Other disease-support groups operate online. Alzheimer's Australia and national DBMAS services also offer a number of additional post-diagnostic support services. See Module 5 – *Managing the impacts of frontotemporal dementia* for further details regarding carer resources.

Brain donation

Obtaining brain tissue from FTD patients after death is a sensitive and important issue. The development of possible treatments depends on understanding the pathology of the illness, which is only facilitated by examination of affected brain tissue. It is recommended that this delicate issue is discussed with families well in advance of the patient's passing.

Clinical experience suggests that families often find tissue donation to be a positive way to contribute to a possible cure for this illness. Specialist referral centres are best positioned to facilitate the brain tissue donation process. A helpful information sheet regarding brain donation has also been developed by Alzheimer's Australia (http://www.alzheimers.org.au/understanding-dementia/update-sheets.aspx).

Summary

Diagnosing FTD can be a complex process which is facilitated by the input of a variety of healthcare specialists. The most important diagnostic information is a clear history of the nature, onset, and progression of symptoms. This information should be collected via independent interviews with the caregiver(s) and, when indicated, with the patient. A thorough cognitive assessment and neuroimaging are also critical to the diagnostic process. It is important to use cognitive and behavioural screening tools which are sensitive to the specific changes displayed by individuals with FTD, as many tools may only detect changes in other forms of dementia. A number of centres in Australia now specialise in the diagnosis of FTD and can provide assistance when this condition is queried.

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