Frontotemporal dementia RESEARCH

Neuroscience Research Australia
Discover. Conquer. Cure.

Each year, one in five people struggle with a major brain or nervous system disorder such as dementia, bipolar, schizophrenia, stroke or Parkinson's, all with life-shattering consequences.

As an internationally recognised research leader, Neuroscience Research Australia's vision is to prevent and cure these diseases and disabilities.

Medical research is the cornerstone of our efforts to advance health and wellbeing for our families and our community.

For many affected by devastating diseases, medical research offers the only hope.

WHAT WE KNOW ABOUT FRONTOTEMPORAL DEMENTIA Dementia is a general diagnostic label that refers to any brain disorder that causes progressive decline in cognitive abilities (memory, language, thinking etc).

There are many causes of dementia. Although dementia is usually considered to be a disorder of the elderly, there is increasing awareness of dementia in younger people (usually defined as onset under 65). Alzheimer's disease is the most common in all age groups, but frontotemporal dementia is the second most common cause especially in younger people. Patients with frontotemporal dementia present with changes in personality and social conduct or decline in language skills known as aphasia. Each new case represents a human tragedy with major implications for the patient, family and friends.

ABOUT OUR RESEARCH The FRONTIER group at the Institute led by Professor John Hodges, Dr Olivier Piguet and Dr. Michael Hornberger is working on a range of topics related to frontotemporal dementia (FTD). Our work focuses on improving early diagnosis, increasing our understanding of how frontotemporal dementia affects memory, language, emotions and problem solving. We collaborate with the genetics group to identify genetic defects that result in FTD and the neuropathology group to understand the cause of cell death in the brain.

WHAT WE HAVE DISCOVERED We know that patients with FTD have problems interpreting the emotion state and thoughts of other people and regulating their own behaviour. We also know that certain aspects of language, notably understanding and word meaning can break down very selectively. We have also begun to understand better which areas of the brain are affected in FTD and how these differ from the changes that occur in Alzheimer's disease. We have also discovered that a range of protein abnormalities result in cell death and that in some instances the cause is a genetic defect.

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Jan Woodmore cares for her husband, Richard, who has FTD. She is a member of the FTD Carer's Group at NeuRA

HOW YOUR SUPPORT HELPS

We are able to make advances in our understanding of the brain and nervous system due to the dedication and generosity of countless people who come to Neuroscience Research Australia every day - research participants, families, carers and supporters.

Donations are an important source of support for medical research – an area with the potential to transform the lives of all Australians through medical breakthroughs.

Your donation or bequest play a key role in allowing us to continue our important research because, ultimately, you are helping our innovative and dedicated researchers to tackle the health challenges facing all of us. For further information visit our website:

www.neura.edu.au

CURRENT PROJECTS

DEFICITS IN EMOTION PROCESSING AND AUTOBIOGRAPHICAL MEMORY IN EARLY FRONTOTEMPORAL DEMENTIA AND THEIR IMPACT ON CARERS

How is the processing of emotion impaired in FTD? How does it affect the ability to remember meaningful and important information from one's life? How does it affect interpersonal relationships? How do these deficits evolve with time? These are some of the questions we are trying to answer. This project is following patients with FTD using tests of emotion detection from static and moving images of faces as well as tests of autobiographical memory. We also interview family members to understand the impact of the disease on interpersonal relationships with measures of carer burden and stress.

BRAIN IMAGING IN FTD Frontotemporal dementia predominantly affects two regions of the brain: the frontal and temporal lobes. Presence of progressive change to these brain structures is an important feature of the disease and helps towards an accurate diagnosis. At NeuRA we have state-of-the-art, high definition brain scan facilities to measure structural changes in the brain. We are studying the extent and severity of changes taking place in the frontal and temporal lobes as the disease progresses. This project will contribute to our understanding of the progression of the disease and improve diagnostic accuracy.

GENETIC STUDIES It is not uncommon for patients with FTD to have one relative with another dementia. In this situation, the risk of dementia for the rest of the family is only slightly higher than the general population. However, a small proportion of patients may have two or more relatives with FTD. This is often caused by a single gene mistake which has been passed on from one generation to the next. These mistakes most commonly occur in the Tau or the Progranulin gene. In addition to laboratory studies on the Tau and Progranulin genes, we are also looking for new genes responsible for families with FTD and motor neuron disease. We hope these studies will allow a better understanding of the biology of FTD and treatment.

SEVERITY RATING SCALES IN DEMENTIA As time passes, FTD can create progressive difficulties in patients. For instance, the ability to perform various activities and in communicating can change. To date, there is not a standardised way to measure change over time in FTD. We have developed an instrument which will help measure such changes. This rating scale will allow professionals to give better advice to families and patients, and make adjustments to their living arrangements.

IMPROVING COMMUNICATION IN PROGRESSIVE APHASIA Patients with the language variants of FTD, often referred to as primary progressive aphasia, lose the ability to communicate effectively with family and friends. We are developing methods of enhancing communication and improving language skill in patients with progressive aphasia

IDENTIFYING BIOMARKERS IN FTD In order to develop treatments and effective interventions for patients with FTD, the underlying process of brain degeneration needs to be identified early in the disease course. FTD patients may have one of several brain pathologies which cannot at present be predicted in life. Cellular studies of the brain have shown that there are two types of protein ("tau" and "Tar DNA binding protein 43") which accumulate in neurons in FTD. The goal of this study is to develop an easily identifiable biological marker for the type of cellular changes occurring in FTD.

OVERLAP WITH MOTOR NEURON DISEASE A proportion of patients with FTD develop features of motor neuron disease (MND), and vice versa. We are investigating the overlap between FTD and MND using a range of cognitive and imaging approaches. We have completed a survey of patients throughout New South Wales supported by MND Australia using instruments that assess behavioural change. We are now testing a cohort of patients attending the MND clinic to examine changes in emotion processing and executive function. In parallel with this approach, we are looking for evidence of motor tract degeneration in patients with FTD using the method of transcranial magnetic stimulation (TMS) for evidence of motor tract degeneration in patients with FTD using TMS.

For further information on supporting research at Neuroscience Research Australia, please call 1300 888 019 or email foundation@neura.edu.au.

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