



19th August 2009

Frontier Update—Referral Criteria Attached

FRONTIER, the first research group dedicated to frontotemporal dementia (FTD) in Australia, was founded in November 2007 with the appointment of Professor John Hodges as Professor of Cognitive neurology at Prince of Wales Medical Research Institute (POWMRI).

We have now assessed over 100 FTD patients, half of whom have a form of progressive aphasia and half have the behavioural variant of FTD (bvFTD). We have also evaluated a number of patients with related conditions such as corticobasal degeneration and FTD associated with motor neurone disease. In addition to the clinical investigations, most of these patients have received a brain MRI as part of our imaging programme. The majority of patients have come from New South Wales, but we have also seen people from Queensland, Victoria, Western Australia and even New Zealand. We have run three successful carer meetings which have seen the initiation of a carer support group and have also organised two training days for professionals in the field. We are very proud of our website (<http://www.ftdrg.org>). This website, which is designed for both professionals and patients and their families, has received very positive feedback.

We are extremely grateful for your support and for referring patients to our programme and to date, we have tried to accept all referrals. Each patient that we see, however, consumes considerable resources to the point that the waiting time for new referrals is now 3-4 months.

It is important to point out that we are funded by the ARC, the NHMRC and the OSMR to undertake research rather than to provide a clinical service but we endeavour to provide information and support for the families that we see who are coping with these disorders. In some instances, however, patients and their families have come with unrealistic or false expectations which we want to avoid in the future.

Now that FRONTIER is out of its infancy, we think it would be helpful to outline clearer guidelines for referrals. These guidelines, as well as our assessment protocol and current research projects are outlined on the attached flyer. Because of the nature of our service and our limited resources, we hope that you will understand if we deem some referrals unsuitable for our projects.



Frontotemporal Dementia Research Group

Criteria for referral

1. A diagnosis of suspected early FTD including bvFTD, Semantic Dementia, Progressive Non Fluent Aphasia and Corticobasal Degeneration established by a specialist (neurologist, geriatrician, psychiatrist);
2. These individuals will be in the relatively early stages of the disease to allow for complete cognitive assessment. We have not found it useful to evaluate patients with marked cognitive, language or physical disabilities. Indications of early stage disease will include one or more of the following:
 - still living at home;
 - still able to communicate if aphasic;
 - preservation of basic self-care activities and continence;
 - able to walk unaided;
3. High-level proficiency in English;
4. Absence of any major confounding neurological or psychological disorders such as any major stroke, severe brain injury, schizophrenia or bipolar disorder.

Our Assessment Protocol includes

Prior to the visit

1. Collection of relevant clinical assessment information;
2. Mail out of behavioural, activities of daily living and life history questionnaires to patients and their families.

At POWMRI

1. Clinical interview by Professor John Hodges with patient and family members;
2. Assessment of activities of daily living proficiency. Typically, this assessment takes two sessions each of approximately 2 hours;
3. Cognitive, language and emotion assessments. This typically takes 2-3 sessions;
4. Brain MRI scan on 3T scanner;
5. Blood sample taken for biomarkers and possible DNA analysis.

If more convenient, some of the assessments can also be conducted in the patient's home.

Current Research Projects

Deficits in emotion processing and autobiographical memory in early FTD

Changes in emotion and empathy are commonly reported in FTD. We are using novel tests to assess the detection of emotions, how these relate to autobiographical memory, the impact on carer's well-being and disease evolution. This NHMRC funded project will follow patients with FTD who undergo tests of emotion, problem solving memory, language and thinking abilities plus a brain MRI scan at yearly intervals.

Brain imaging in frontotemporal dementia

At the POWMRI we have modern, high definition brain scan facilities available to measure structural changes in the brain. In this project, we are studying the extent and severity of changes in the frontal and temporal lobes as the disease progresses.

Severity rating scales in dementia

To date, there is not a standardised way to stage dementia in FTD and to measure change over time in FTD. We are developing an instrument which will help measure such changes and allow professionals to give better advice to families and patients.

Assessing and improving communication in progressive aphasia

Patients with the language variants of FTD often, referred to as primary progressive aphasia (PPA), lose the ability to communicate effectively. We have been developing a new battery to evaluate and sub-classify patients with PPA. In conjunction with Speech and Language Therapists in Sydney, we are hoping to develop methods of enhancing communication and improving language skill.

Identifying biomarkers in frontotemporal dementia

In order to develop treatments and effective interventions for patients with FTD, the underlying process of brain degeneration needs to be identified in vivo. Recent neuropathological studies have shown that abnormalities of two different proteins ("tau" and "Tau DNA binding protein 43") have a key role in the causation of FTD. The goal of this study is to develop blood-based biological markers for tau, TDP43 and other pathologies.

Overlap with motor neurone disease

A proportion of patients with FTD develop features of motor neuron disease (MND, also known as Amyotrophic Lateral Sclerosis) and vice versa. We are investigating the overlap between FTD and MND using a range of neuropsychological, and imaging tools. As part of this study patients with MND undergo a motor examination and brain TMS to look at central motor conduction.

For further information, please contact: Professor John Hodges via email on j.hodges@powmri.edu.au or his Personal Assistant, Sarah Homewood, on 9399 1134 or email FRONTIER@powmri.edu.au