

Helping the FTD Patient-Caregiver Dyad

Can. J. Neurol. Sci. 2011; 38: 671-672

"To cure sometimes, to relieve often, to comfort always."

— Edward Livingston Trudeau

Like Alzheimer's disease (AD), Picks disease, or its contemporary incarnation Frontotemporal dementia (FTD)¹, is a clinical neurodegenerative syndrome of insidious onset that ultimately strips the afflicted of all cognitive and functional independence and inexorably leads to death, usually within a decade of initial detectable changes. Although less common across age groups, it's prevalence is equal to that of AD in individuals under that age of 65, with onset usually between 45 and 65 years^{2,3}.

Despite preceding the description of AD by nearly a decade, FTD has remained in the shadows of its younger cousin, both from a clinical and a scientific point of view. But, academic interest in FTD is growing, as reflected by the number of PubMed publications on the topic. Peaks in publication occurred following consensus meetings in 1994⁴ and 1998⁵, and more recently following the identification of the elusive ubiquitinated protein inclusions TDP-43^{6,7} and FUS⁸, as well as the role of progranulin mutations^{9,10}. In fact, our exploding understanding of the molecular neuropathological underpinnings and their correlation to the clinical manifestations of this heterogeneous group of disorders is also leading to the isolation of more pathologically homogeneous groups during life¹¹ and the appearance of clinical trials designed to address etiology¹². Although current therapies are limited to symptomatic palliation¹³, successful disease modifying treatment will likely emerge within our lifetimes.

Patients with FTD have also contributed selflessly to the advancement of our understanding of brain-behavior correlations: Application of modern brain imaging techniques to these patients has uncovered the neuroanatomical substrates of complex concepts such as empathy, recognition of emotions and emotional blunting¹⁴, aberrant motor behavior and stereotypies¹⁵, apathy¹⁵, disinhibition¹⁵, binge eating¹⁶, self appraisal¹⁷, and even interpersonal traits¹⁸. Cognitive and behavioral neuroscience has made giant strides in our understanding of what is it to be human thanks to the study of patients with this disorder.

But what are we doing for these patients now? What do these patients and their caregivers actually need the most? Frontotemporal dementia strikes in the prime of life. Patients are often fathers or mothers of young families and in the midst of their professional careers. The social, professional, and even legal consequences of the disorder are frequently devastating and, unlike those of the later onset Alzheimer's disease, have an impact that reaches far beyond the individual patient.

As is often the case with chronic diseases, patients and – especially for dementias – caregivers will be the impetus for the creation of support groups and ultimately associations like the Alzheimer's Society, and for FTD, the Association for the FTDS

(www.theaftd.org), created in 2002. These groups, and numerous other players involved in the diagnosis, management, and study of patients with FTD address ongoing issues that occur once diagnosis has been made.

In their paper in this edition of the CJNS¹⁹, Tiffany Chow and her colleagues from the University of Toronto and Dalhousie University, have basically asked their patients' caregivers "how are we doing?" Their questions were simple (and paraphrased here): In addition to probing for the initial manifestations of the disease with "What were the first signs of disease?", they explored the impact of the disease and of the diagnostic process on their lives with "What frustrates or frustrated you most?", "Who or what helped you most?", and "What surprised you the most?" Through an innovative mechanism using an online survey technique, they were able to obtain candid opinions and responses to their questions from caregivers from Canada, the US, Britain, and Europe. With all the caveats associated with voluntary online surveys, the results of their study are rather telling.

Concerning initial changes, the most frequent was "change in thinking and judgment", which included "insensitivity". The most troublesome aspects of caregiving included "poor insight", "caregiver quality of life". What surprised caregivers the most was that "dementia can affect young people". Finally, resources or interventions that help the most are striking: "the treating neurologist" came first, followed by "the Internet".

In isolating the treating neurologist and the Internet as the two most helpful resources, their data betrays the prolonged frustration experienced by the caregivers in getting the correct diagnosis and the appropriate guidance and help. The problem is, FTD is hard to recognize early, and hard to diagnose. Families are aware that something is awry long, long before a correct diagnosis is suggested. In contrast to AD, where the lead-time to diagnosis can be up to three years after appearance of noticeable changes, patients with FTD can progress for five or more years before diagnostic efforts begin to focus on this etiology. Although caregivers of patients with FTD may wait twice as long as caregivers of patients with AD before seeking medical advice, physicians will take nearly an extra year to actually make the diagnosis. In addition, twice as many patients with FTD are likely to receive initial non-dementia diagnoses, including psychiatric conditions²⁰.

Indeed, if we are going to help the caregivers and this patient population (and in particular if we hope to get them involved in early etiological treatment trials), we need to educate the public about the existence of "dementia in the not-so-old". Further, in an effort to shorten the delay in seeking medical advice, families need to understand and recognize the earliest changes associated with this group of disorders²¹. This study clearly identifies the Internet as a resource for supplying appropriate information to those unwitting caregivers reaching out and grasping to understand what is happening to their loved one.

Any delay to seeking medical advice notwithstanding, the onus really falls upon us to educate ourselves. We need to be able to make the diagnosis earlier and refer to specialized tertiary care centers where appropriate interventions such as clinical trials, specialized daycare, and disease-adapted legal counseling can be obtained²²⁻²⁴. Because studies such as this one also capture the caregivers' first appreciation of change, they help to translate caregiver complaints into a differential diagnosis that must include FTD.

Clearly, further studies of this kind will help bridge the gap between what both families and doctors know and what we need to know, not only to better comfort, but, ultimately, to cure this devastating disorder.

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