



Frontotemporal dementia and psychiatry

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EDITORIAL

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I wish it would dawn upon engineers that, in order to be an engineer, it is not enough to be an engineer. (Jose Ortega y Gasset, *Toward a Philosophy of History*, 1941)

Ortega y Gasset argued that a profession immersed in the everyday routines of its business faced insularity and entrapment within its boundaries. Therefore, a profession, and its professionals, must be watchful of “the entire scene of life”, i.e. they must be alert to the dynamic interplay between current circumstances and new developments. All psychiatrists ought to be familiar with frontotemporal dementia (FTD). It was the psychiatrists Arnold Pick and Paul Sérieux who, more than 100 years ago, provided the original clinical descriptions of frontotemporal dementia (Pick, 1892; 1905; Sérieux, 1893). Aloisius Alzheimer, another psychiatrist, provided the seminal description of its neuropathology (Alzheimer, 1911). Psychiatry’s interest in neurophysiology and neuropathology declined dramatically after the 1930s; dementia and FTD research was rekindled in the late 1960s (Brun, 2007) after a decades-long interregnum imposed by the psychoanalytic era. The paradox is that while psychiatry has readily re-engaged in the treatment and study of Alzheimer’s disease, the profession still shows comparatively little interest in FTD and other non-Alzheimer primary dementias. Yet judging from its clinical features, FTD is a natural subject for psychiatric interest and inquiry. This disorder is usually characterized by progressive decline in conduct and socialization consisting of inexorable coarsening of temperament, dispositions, judgement and comportment, decline in emotional functions and self-management, and disintegration of speech and communication. In other words, the classic syndrome is primarily a behavioural disorder – although there is diversity in the phenotype. Furthermore, FTD causes suffering in families that may manifest as depression, anxiety, sleep disorders and other psychiatric states that may arise during stress.

Our participation in the treatment and study of FTD has potential benefits for psychiatric knowledge and practice. For example, studies of social cognition in FTD have provided key insights into the neurophysiological and neuroanatomical substrates of emotion recognition, interpersonal behaviour and social reasoning, particularly for constructs such as empathy (Rankin et al., 2006) and extraversion (Sollberger et al., 2012, 2009). FTD patients often develop parts of psychiatric syndromes rather than the whole, such as the apathy and anhedonia seen in major depressive disorder without sadness or loss of self-worth. Thus FTD provides an opportunity to understand the neuroanatomy of components of psychiatric syndromes. FTD research is also beginning to provide insights relevant to eating disorders (Piguet et al., 2011; Woolley et al., 2007). In turn, psychiatry offers much to FTD treatment and research. Psychiatric expertise in the examination of mental states and dysfunctional behaviour has contributed to our understanding of FTD phenomenology (Brun, 2007), and the treatment of FTD frequently relies on the management of offensive and maladaptive behaviours in psychiatric ambulatory settings and hospital wards. Psychotherapeutic agents are frequently prescribed during FTD care (Hu et al., 2010), and psychiatric resources for psychosocial adaptation offer creative and effective delivery of various rehabilitative and support services to alleviate the suffering of the patients and their carers.

This issue of the journal aims to reintroduce psychiatry and psychiatrists to FTD. Papers in this issue describe FTD and its clinical manifestations, review how the diagnosis is made, and discuss the latest diagnostic criteria. Other papers explore psychiatric facets of this illness, review psychiatric differential diagnosis, and examine epidemiological and neurobiological connections between FTD and schizophrenia. We have reviews of FTD epidemiology, measurement and diagnostic controversies; and of neuropsychological and physiological markers. The psychiatric and community models for FTD care and neurorehabilitative interventions are also covered.

One paper introduces corticobasal degeneration (CBD) and progressive supranuclear palsy (PSP), two conditions closely related to FTD. We are very grateful to our contributors who, despite busy schedules, have managed to write these excellent papers.

Discoveries in the genetics of hereditary FTD over the last 15 years have markedly accelerated the depth and scope of FTD neurobiology, and there is now active investigation of the pathophysiologic mechanisms for disease. These genetic discoveries also provide occasion and opportunity to refine our knowledge of the phenotype. The clinicopathological studies of patients who have progranulin and chromosome 9 open reading frame 72 (C9ORF72) mutations have provided intriguing observations about the FTD phenotype and its diversity. Psychosis is a common feature in C9ORF72 carriers (Snowden et al., 2012), and onsets with bipolar disorder (Floris et al., 2013) and suicide attempt (Synofzik et al., 2012) have been reported. A recent study did not find C9ORF72 carriers in a large cohort of schizophrenia subjects (Huey et al., 2013), but an earlier study described a family where the progranulin mutation was associated with FTD in one brother and schizophrenia in the other two (Momeni et al., 2010). Thus there are indications that the FTD phenotype is broader in progranulin and C9ORF72 mutation carriers, and includes presentations that mimic primary psychiatric disorders. These observations also suggest that studies of hereditary FTD have the potential to help resolve a paradox of psychiatric genetics: we know that many psychiatric disorders are heritable, yet genome-wide association studies have not identified the major components of this heritability. Examination of highly penetrant mutations that impair conduct and cognition, and cause abnormalities of affect and psychosis, such as those that result in FTD, may help us understand the genetic basis of common psychiatric illnesses.

Psychiatry has a large role to play in the treatment and study of FTD. The hope is this issue will educate and stimulate the reader and, perhaps, inspire a reimagining of the business of psychiatry. New discoveries have rendered professional boundaries defined by “organic” versus “inorganic” status obsolete and are reshaping our understanding of dementia and psychiatric phenotypes. Psychiatry and its practice might in the future be defined by a focus on disorders of mental life and behaviour, together with the skills, perspectives and resources that we offer, without the constraints of arbitrary and superficial pathophysiological boundaries.

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