

GUEST EDITORIAL

Uncommon causes of dementia: rare, but not marginal

Dementia is one of the most frequent and disabling health problems of our times. During the past twenty years, we have witnessed growing public and professional interest in dementia, its diagnosis and causes. This has led to the introduction of better methods of detection of cognitive impairment, coupled with early diagnosis of Alzheimer's disease and, to a lesser degree, of other forms of dementia. However, if the clinical presentation of patients does not follow the typical pattern of progressive memory loss starting in later life, then currently available diagnostic methods are less likely to be useful and patients with dementia may go unidentified for prolonged periods of time. Some of the less frequent causes of dementia may initially present with psychological or behavioral disturbances, others are associated with prominent neurological signs and symptoms. Patients may also present with marked deficits in cognitive domains other than memory, such as impaired language, which makes the diagnostic workup more difficult and onerous.

There is also a risk that difficulties related to the accurate diagnosis and investigation of these conditions may be dismissed as irrelevant by clinicians and researchers alike, simply because they are "uncommon." After all, what is the point in worrying about something that only affects a small proportion of the population? We do not agree for the following reasons. First, many of the uncommon causes of dementia can be improved or even reversed if adequate treatment is introduced promptly (for example, when dementia is associated with certain types of infection; see Almeida and Lautenschlager, in this issue). Second, effective preventative strategies are also possible once the pathogenetic mechanisms are, at least partly, understood. The public health measures introduced to control the spread of the new variant of Creutzfeldt–Jakob disease is but one such an example. Third, a better understanding of the steps that lead to the development of dementia in some of these uncommon conditions may provide valuable information about the pathogenetic processes involved in the causation of conditions such as Alzheimer's disease. For example, most of the currently available knowledge about the genes and proteins associated with Alzheimer's disease have originated from the study of a very small number of families with extremely uncommon genetic mutations that result in the development of Alzheimer's disease with early onset (see Novakovic *et al.*, and Lautenschlager and Martins, this issue). New therapeutic approaches for Alzheimer's disease are currently being developed that take into account this new knowledge.

This supplement was conceived with the aim of renewing people's interest and knowledge about some of the less frequent, yet fascinating, causes of dementia. The papers in this issue cover a variety of topics: historical background and classification, uncommon genetic neurodegenerative disorders, infections, drugs, neurological and metabolic diseases, autoimmune and endocrinological conditions, toxic and psychiatric illnesses. A paper outlining the perspective of carers of patients with dementia completes this special issue of the journal. Together, these papers represent a unique resource for the clinician, covering an array of epidemiological, clinical and pathogenetic aspects of a variety of "non-Alzheimer causes of dementia" that are often listed in textbooks but are hardly ever discussed in detail. We have endeavored at all times to make this supplement both accessible and useful to the busy clinician involved in the care of patients who may present via a myriad of pathways.

We have had the privilege of working alongside a talented and knowledgeable group of colleagues who kindly agreed to set time aside to prepare valuable contributions to this supplement – we are grateful to all of them. We also thank the International Psychogeriatric Association for their support and advice, and Professor David Ames, Editor of *International Psychogeriatrics*, for encouraging and enabling us to publish this special issue of the journal.

We trust that you will enjoy reading the papers in this supplement as much as we did.

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