Preface

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The classification of dementia has been a controversial issue ever since the evolution of modern neuropsychiatry began at the end of the 19th century. At that time, Arnold Pick reported an association between circumscribed cortical degeneration and aphasia and behavioural changes in some cases of dementia and Alois Alzheimer later described the histopathological hallmarks of the clinicopathological entity of Pick's disease. The concept of frontotemporal dementia (FTD) has evolved mainly during the last two decades, when besides the rare Pick's disease also non-Pick forms were discovered and first reported by the Lund Dementia Study Group in Sweden [1] and by the Manchester group in the UK [2]. The spectrum was then widened through the discovery of gene-related forms such as FTD-3 and FTD and Parkinsonism linked to chromosome 17 and other important contributions from research centres all over the world. Stimulated by these developments, the Lund group arranged international conferences on FTD in Lund in 1986, 1992 and 1998 [3–5]. The 4th FTD conference in 2003, in Lund, is here reported.

As in the past, scientific exchange in a new field such as this one may become troubled by nomenclature inconsistencies. Attempts to overcome this have been made on a couple of occasions through clinical and pathological consensus [6, 7]. As to genetics of these disorders as a dividing ground, the rarer forms are known to be caused by various mutations on for example chromosomes 17, 3, and 9, while possible mutations are unknown in the vast majority of FTD cases, most of which are sporadic. Another possible principle for classification would be biomarkers. Lately a classification has been suggested [8]

based on molecular biological findings such as tau or synuclein subforms, dominated by rare forms, whereas again the by far dominating form of FTD yet lacks specific markers for a subdivision. This question was addressed aiming at tau, considering the dominating FTD form to be tau-less. From the standpoint of present knowledge, however, a classification based on tau might attract too many and diverse disorders, given the communality of tau alterations.

Several clinical and experimental papers at the Lund conference 2003 dealt with biomarkers in chromosome 17- and 3-related forms. Hopefully such work will in the future offer a more logical basis for the classification of FTD. Clinical-phenotypical variants of FTD, some related to genetic findings, was another field with several contributions, one pointing out the considerable phenotypic variation between and also within families with the same chromosome 17 mutation and interestingly one form of FTD and Parkinsonism linked to chromosome 17 closely resembled Pick's disease. Other contributions stressed the uniformity among a large number of affected members with the chromosome 3-linked form. The intriguing coupling between amyotrophic lateral sclerosis and FTD was found to be more common than previously reported and a search for responsible mutations is under way.

Epidemiological studies point out that FTD is a more common form of presenile dementia than previously recognized. Neuroimaging showed that contrary to some earlier results PET hypometabolism in semantic dementia is initially restricted to the temporal lobes compared with other FTD forms which show a frontal decrement, in

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addition to lateralization. This means that these forms start focally and asymmetrically, later to diffuse and become widespread and rather symmetrical in distribution as seen at autopsy.

The development of therapies for FTD was dealt with in several presentations. Specific disease-modifying treatment is unavailable, but positive experience with symptomatic pharmacological interventions was reported. There were also presentations of the announcement of the Association for Frontotemporal Dementias in the USA, the establishment of the Swedish National Competence Centre for FTD in Lund and facilities to support the care for FTD by the Swedish Alzheimer Association.

After four conferences on the theme in Lund, Sweden, the relay baton was handed over to Murray Grossman, Bruce Miller, Virginia M-Y Lee, John Trojanowski and

John van Swieten to arrange in 2004 the next conference in Philadelphia as a satellite meeting at the 9th International Conference on Alzheimer's Disease and Related Disorders, July 2004.

The 4th International Conference on Frontotemporal Dementia was arranged by the Lund Dementia Study Group, University of Lund, Sweden, in cooperation with leading research groups from all continents and sponsored by the Alzheimer Foundation, Sweden, The Crafoord Foundation, Sweden, Ellen and Henrik Sjöbring Foundation, Sweden, Konsul Thure Carlssons Minne, Sweden, Jansen-Cilag AB, Sweden and France, Ratiopharm AB, Sweden, William and Py Gibson Foundation, Sweden, H Lundbeck AB, Sweden, Merck, Sharp and Dome, Novartis Sweden AB, Pfizer AB, Sweden and ADERMA, France.

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