Global democratic consensus on neuropathological disease criteria

On July 10 and 11, 2002, a group of experts in Parkinson's disease attended a satellite meeting of the 7th European Congress of Neuropathology in London, UK, and agreed on a core definition of the histopathological phenotype of Parkinson's disease, which takes into account recent developments in the field (eg, α-synuclein immunohistochemistry) but essentially abandons many conventional criteria as they are no longer sufficient. This definition and comments can be viewed at http://www.ICDNS.org.

It rapidly became clear during the discussions at this meeting—and subsequently at the European Congress in Helsinki, Finland (July 13–16, 2002)—that a thoroughly democratic voting system on disease criteria, which uses the worldwide web to provide global access to all qualified diagnosticians, would represent a fundamental breakthrough. The plan was adopted, therefore, to publish diagnostic criteria for all recognised neuropathological diseases on the web, where the global community of neuropathologists can judge them. Acceptance should be facilitated greatly by the fact that no named individual or national group is leading the initiative. Instead, existing classifications will be "translated" into a generic format avoiding personal as well as institutional names to ensure consistency of terminology between related disease processes.

The majority of neurological diseases are currently defined on the basis of their histopathological phenotype. However, molecular definitions are likely to apply even to common, genetically heterogeneous, or "complex" neurological disorders, such as Parkinson's and Alzheimer's disease. Molecular definitions are advantageous as they provide a rationale for precisely targeted, individualised therapies that are most cost effective.

Degenerative diseases are among the most common neurological disorders—many are related to ageing and are late in onset. Consequently, it is expected that the prevalence of these diseases will rise significantly over the next two decades in the western population, followed by a similar, although less dramatic, increase in other populations.

An important development with regard to disease classification concerns the increasingly quantitative nature of biomedical research. Although the single hypothesis-driven scientific approach has been widely used during the past few decades, "exploration science" is gaining predominance with the introduction of high-throughput analysis methods based on microarrays. This approach is employed widely in genomic, transcriptomic, and proteomic research and is beginning to be used in clinical settings.

Quantitative definitions of histopathological phenotypes could be linked to array data sets and combined with imaging and clinical parameters to produce a multidimensional data space from which binary codes representing "signatures of disease" can be extracted. The implementation of these exciting new opportunities into the daily practice of neurologists and other clinical neuroscientists will benefit greatly from global consensus on key criteria that are used globally for making a histophenotypical diagnosis.

However, it is surprising that an international consensus on neuropathological disease criteria does not exist, except in the field of brain tumours where the WHO backs an existing histological classification. Consequently, the WHO has been asked to do the same for the neurodegenerative diseases group but the problem clearly extends to other diseases of the nervous system as well.

The proposed way of developing such criteria is for recognised experts to prepare core definitions that are published anonymously online. Diagnosticians can then suggest modifications, which are signed and dated. All users have to register to confirm their identity. Each comment—if not withdrawn by the author before an agreed deadline—will become part of the history of a disease definition. No opinion will be ignored provided it comes from a qualified colleague as confirmed by the respective disease "moderator". When a consensus has been reached, a vote will be taken to accept a published version of a classification or diagnostic criteria. Both core definitions and comments can be used immediately for diagnostic purposes as outlined in the guidance section of the website (http://www.ICDNS.org).

Core definitions are expected to receive both positive and negative votes. Although the details remain to be worked out, the immediate advantage of such a scheme is that core definitions can be updated in line with developments in the field either by expert groups (eg, during specialist meetings) or individuals—both have to be approved by a democratic poll on the web. This concept was discussed and backed by the signatories below. Terms of reference have been posted and are now open for discussion at http://www.ICDNS.org.

For the first time, a global consensus on histopathological diagnostic criteria is within reach. Neighbouring professional disciplines, such as neurology, neurosurgery, neuroradiology, and psychiatry, are strongly encouraged to engage in similar activities. This should result in a common basis for correlative molecular work which will be of utmost importance in the future.

Cosignatories (in alphabetical order)

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