

# Book Reviews

## **Disability and Culture: Universalism and Diversity**

Edited by T Bedirhan Üstün

Seattle, Hogrefe & Huber/World Health Organization. 2001, pp 328, £31.45, US\$ 44.50 (Paperback)  
ISBN 0 88937 239 X

This volume describes the cross-cultural applicability research (CAR) study undertaken as part of the revision of the World Health Organization's International Classification of Functioning and Disability (ICIDH-2; 2001) first published by WHO in 1980. The ICIDH-2 has done more than update vocabulary (to Body function and structure, Activity and Participation). It is based on the understanding that disability is a complex phenomenon, arising from one or more health conditions manifested at the body, person, or social levels, and affected by contextual factors such as environment and personal choice. It thus incorporates both the medical and social models of disability. As a measurement tool, the ICIDH-2 is intended to be useful in measuring the impact not only of physical and sensory impairments, but also of mental health conditions and alcohol and drug related conditions, given the major burden of such disorders on society. Fundamentally, disability is not seen as a defining feature of a subset of persons, but, rather, a universal condition of humanity itself.

The CAR study was conducted in 18 centres, including India, Canada, Turkey, Nigeria, Spain, UK, and Japan. The aims were to ensure that the ICIDH-2 '...concepts, and the [disability assessment] instruments, could be used to compare international disability statistics correctly and in planning comparable and effective health services around the world' (p 21) – a tall order. Assessment requirements included the need for cross-cultural comparison of terms, identification of anchors that would determine thresholds for disability and severity (e.g. subjective distress, how noticeable dysfunctioning is), and identification of positive and negative attitudes in different cultures. The 18 centres were asked to collect extensive information on translation and linguistic analysis, pile sorting of terms included in the classification, concept mapping, key informant interviews, and focus groups. Each centre had to recruit groups of people to undertake these tasks, including medical or other health professionals, persons with disabilities, carers, and policy makers.

From the start, it is clear that the process was different in each of the centres. The views obtained usually represented opinions of those known to the centre investigators (mostly urban), and local knowledge of disability matters was often fragmented. Given the range of health conditions covered in the exercise, this is hardly surprising but it means that the representativeness of the opinions gathered cannot be judged adequately.

Individual chapters from each of the centres make interesting reading. For example, in Cambodia almost all the contributors to the process had physical disability or worked in physical rehabilitation, often connected with amputations (from mine explosions) or polio. Yet, public attitudes were described as generally negative, with individuals with dis-

abilities being seen as morally at fault and were, thus, excluded from social activities and normal roles. In some other countries, the family was seen as being responsible for the disability, or the individual assumed to have transgressed in their 'past life'. In other cultures, however, people are treated more sympathetically when the disability is seen as arising from a birth defect or an accident, than if it is seen as 'self-inflicted' as in the case of drug and alcohol-related conditions. Attitudes have an effect upon the degree of public assistance available to individuals and families.

Difficulties in translation also make interesting reading, with several languages evidencing problems with crucial terms such as 'affect', 'community', 'disability', 'disablement', 'disorder', 'function', and 'well-being'. Possible solutions are suggested, but there is a recognition that parts of the classification system may not be culturally applicable in a small number of cultures.

This book is not an easy read. It is a research report, consisting of multi-authored edited chapters, with significant overlap and repetition. The focus of the CAR study was very broad, and entirely adult-oriented. For example, the key informants were asked about disability thresholds in relation to five scenarios, describing in turn an adult with a mobility problem, mental disorder, low intelligence, alcohol problem, or heroin problem. This was hardly likely to result in testing of conceptual frameworks of relevance to developmental disability. Indeed, the country reports frequently seemed impossibly positive about the situation for people with physical or sensory disability; many countries reported that, out of a list of 18 health or economic conditions, social reactions would be most positive and stigma least for individuals in wheelchairs, or blind people. This raises the question of whether the whole exercise was skewed by the inclusion of drug and alcohol problems. As a detailed example of a complex, cross-cultural, qualitative study, the book has its merits and uses. But it won't be a best-seller.

*Helen McConachie*

**West Syndrome and Other Infantile Epileptic Encephalopathies**  
Guest Editor Y Fukuyama. *Brain and Development: Special Issue*. 2001; 23: 441–778.

It is a commendable achievement by Fukuyama and his co-authors to have published the proceedings of the International Symposium on West Syndrome and Other Infantile Epileptic Encephalopathies so soon after the event. The result is a combination of review and original articles. However, many of the contributions that are designated as reviews contain information that is probably not widely known to clinicians. Examples are: findings from the use of newly developed tracers in PET, discussions on the corticotrophin releasing hormone excess theory, and the possible relevance of the central histaminergic neuron system as an anticonvulsant mechanism.

Several authors consider the special relationship between the onset of spasms and maturational age. Specific anatomical, neurophysiological, and biochemical features of the brain in the postneonatal and early infantile periods are reviewed in this context. An interesting original article

describes serial EEGs in infants with neonatal hypoxic ischaemic encephalopathies who are, therefore, at risk of developing West Syndrome (WS). They showed that for those who were born preterm, there was a period before the development of hypsarrhythmia, when isolated epileptiform discharges were recorded. On the other hand, those born at term developed hypsarrhythmia without an earlier, less severe stage. Dulac, in a paper entitled 'What is West Syndrome?', discusses the inclusion of patients with wider than usual ages at onset and atypical clinical and EEG patterns, but clearly excludes those in whom early myoclonic encephalopathy and early infantile epileptic encephalopathy (EIEE, Ohtahara syndrome) would be more appropriate diagnoses. Several authors question the decision of the Commission on Classification of the International League against Epilepsy to define WS as a generalized epilepsy. Evidence from neuroimaging, as well as from clinical observation supported by video-recording, seems to favour the concept that in the majority of cases of WS the spasms are associated with focal brain abnormalities.

Of the original articles, several relate to national surveys, epidemiological studies, or hospital-based reviews of WS. Most come from the countries of Southern Asia.

Methodology, period of ascertainment, length of follow-up, and information on treatment vary somewhat from study to study. Nevertheless, they undoubtedly contribute to overall knowledge of this relatively infrequent condition, and could be the beginnings of a multinational data base. A paper on familial cases is a reminder that some cases of WS are genetically determined. The association of WS with tuberous sclerosis complex is considered in both review and original articles. Treatment with ACTH, vigabatrin, high-dose vitamin B6, zonisamide and thyroid-releasing hormone is reviewed. However, the available evidence does not allow absolutely clear guidelines on therapy to emerge. It is disappointing that although topiramate is reported to be used increasingly in Korea, its efficacy is not specifically considered. The contributions on prognosis emphasize the continuing disabilities of most patients who present with WS, but identify the possibility that those with tuberous sclerosis may do better than others with identifiable lesions, particularly if they are treated with vigabatrin.

Considering the cohesion of the papers on WS, those on other epileptic encephalopathies of infancy seem rather less focused on a general theme. However, it is useful to have more information on the clinical, EEG, and neuropathological aspects of EIEE with suppression-bursts; and to have the importance of non-ketotic hyperglycinemia as a cause of early myoclonic encephalopathy re-emphasized. A single case of surgical treatment of EIEE is reported.

A contribution on refractory grand mal seizures with onset at infancy draws attention to the difficulty with accommodating all infantile seizure disorders within the currently accepted syndromal definitions. WS and the other epileptic encephalopathies of infancy remain some of the most challenging of the epilepsies of infancy and childhood. Both those who deal with the acute presenting phases and those

who look after the children with longer-term disabilities will find much of relevance in this special issue.

*Sheila J Wallace*

### **The Cerebellum and its Disorders**

Edited by Mario-Ubaldo Manto and Massimo Pandolfo  
Cambridge, UK, Cambridge University Press. 2001, pp 589  
£150.00, US\$225.00  
ISBN 0 521 771156 0 (Hardback)

The editors of this 572-page volume are to be congratulated for bravely taking up and completing the enormous task of reviewing the multiple aspects of cerebellar development, structure, functions, and disorders. Most of the 70 contributors are acknowledged authorities in the field.

The first nine chapters of the book include extensive coverage of what is known about embryology, anatomy, structure and function of the cerebellum, and all these chapters are of very high quality. Chapters on the theories of cerebellar control and the correlation of clinical signs to physiological dysfunction are especially interesting and helpful. The chapter on the role of cerebellum in cognition and affective functions is of great interest. It goes well beyond the relations of cerebellum with psychosis as stated in its title, and covers, superbly, a field that is currently the subject of much speculation and study.

As in all multi-authored texts, the coverage is uneven; some chapters are excellent up-to-date reviews while others offer somewhat scanty coverage.

Some difficult issues (e.g. the relationship between the classifications of olivo-ponto-cerebellar atrophies and spinocerebellar atrophies) would benefit from some discussion as they are far from clear in many neurologists' minds.

The reasoning behind the choice of disorders selected is somewhat mysterious. For example, diseases in which cerebellar involvement is not a prominent feature, such as corticobasal degeneration or the Shy-Drager type of progressive multiple atrophies, are well covered, whereas conditions like the leukodystrophies or the myoclonic epilepsies (with the exception of dentato-rubral, pallidolusian atrophy) are ignored, as are disorders of amino acids and organic acids metabolism. Refsum's disease, abetalipoproteinemia, ceroid lipofuscinoses, the early-onset recessive ataxias, and vitamin deficiencies such as vitamin E are only briefly dealt with, with few (if any) references.

This book, however, constitutes a major reference text for those primarily interested in the basic aspects of cerebellar functions and dysfunctions. It brings information in this domain up to date in a clear and lucid fashion. Many of its excellent chapters will be of interest to clinical neurologists.

Child neurologists will appreciate the high quality of the basic information contained in the volume. I fear, however, they may be disappointed by not finding sufficient coverage of many of the diseases they struggle with in their practice.

*Jean Aicardi*