

Book Reviews

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London Dysmorphology Database, London Neurogenetics Database & Dysmorphology Photo Library on CD-ROM. 3rd ed. By Michael Baraitser and Robin M. Winter. Oxford: Oxford University Press, 2001. \$2,495.00 (Windows CD-ROM).

First published, in 1990, as the *London Dysmorphology Database* (with a separate neurogenetics database released the following year), the 3rd edition, published as the *London Dysmorphology Database, London Neurogenetics Database & Dysmorphology Photo Library on CD-ROM*, combines two comprehensive databases, together with an extensive photo library and free online updates for registered purchasers, onto a single CD-ROM. An accompanying 75-page instruction manual makes the software easy to install, and its eight chapters are replete with examples and illustrations (including a glossary of electrophysiological terms) that should enable even the most computer-challenged user to derive the maximum benefit from the two databases and photo library. Switching between databases is easily accomplished, and links to Online Mendelian Inheritance in Man (OMIM) and Medline are additional features.

Dr. Baraitser and Prof. Winter have included more than 3,400 nonchromosomal syndromes (with their OMIM accession numbers and chromosomal loci, when known) in the dysmorphology database and nearly 3,300 neurological disorders in the neurogenetics database—along with more than 12,700 thumbnail photographs that, with a double click of the mouse (when the CD-ROM is inserted), can be displayed in detail. By use of the “My Collection” function, it is possible to edit photograph captions in the database and to import images from one’s own personal collection, as well.

Every work, even an effort as encyclopedic in scope as the *London* databases, can be further improved. In the neurogenetics database’s syndrome keyword search, for example, to find an entry for oculopharyngeal dystrophy, the user first has to look under “muscular dystrophy”—“muscular dystrophy - oculopharyngeal (autosomal dominant)” or “muscular dystrophy - oculopharyngeal - autosomal recessive” (note the databases’ inconsistency in use of parentheses and hyphens)—and, for the hyper- or hypokalemic forms of muscular dystrophy, under “periodic paralysis.” One major drawback is that laboratory abnormalities are not included in the syndrome feature search, nor, inexplicably, does the feature search contain a separate general heading for cardiovascular or heart abnormalities. Instead, the user must go first to “thorax” and then to the subheading “heart.” Although aortic incompetence

is mentioned under “heart,” aortic dissection appears under “blood vessels”—and cystic medial necrosis is nowhere to be found, whereas mitral-valve prolapse is subsumed under “mitral incompetence.”

At \$2,495.00 per set—individual volumes can be purchased for \$945.00—most individual practitioners will find the software prohibitively expensive. Nonetheless, metabolic disorders services, neurology clinics, and genetics centers, especially those involved in training programs, should find the databases to be a useful adjunct to standard reference works. Dr. Baraitser and Prof. Winter are to be commended for contributing a valuable resource to the literature of dysmorphology and neurogenetics. I would, however, offer one suggestion to enhance future versions of the *London* databases—namely, the inclusion of a section on chromosomal disorders.

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Disorders of Voluntary Muscle, 7th edition. Edited by George Karpati, David Hilton-Jones, and Robert C. Griggs. Cambridge: Cambridge University Press, 2001. Pp. 775. \$220.

This is a multiauthor book edited by three eminent specialists in the neuromuscular field, who have gathered together an impressive number of outstanding contributors.

What did I like about this book? I think it is a well-balanced combination of fundamental aspects of muscle physiology, investigations of muscle disease, and descriptions of individual conditions, each contributing approximately one-third of the 755 pages. The first part, on muscle physiology and molecular structure, is exceptionally clear, and I would strongly recommend this to anybody, ranging from medical students to neurology trainees to people like me with a strong academic neuromuscular interest.

I have certainly learned new aspects of a variety of relevant systems, ranging from the sarcotubular system to the extracellular matrix. It is not easy to find an up-to-date, comprehensive, and “digestible” review on this subject that is relevant to the clinical neurologist, but this book has achieved it.

The second part, on methods of investigation of muscle disease and of the patient with muscle disease, puts a strong emphasis on the rational and clinically based approach to the diagnosis of neuromuscular conditions. This is also a very enjoyable part of the book, because of its clarity. It covers many of the fundamentals of “getting a diagnosis right,” and it is these fundamentals that are most frequently ignored, leading to misdiagnosis of individual muscle conditions.

The third part, on individual muscle condition, is also enjoyable; it is obviously an area in rapid evolution, but all the

important and key aspects of individual conditions are clearly and rationally presented, with helpful diagnostic hints.

It is the optimal balance between these three parts that makes this book especially valuable for people with an interest in neuromuscular disorders.

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