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Neurofibromatosis Type 1 in Childhood.

Kathryn North, David Gutman, Bruce R Korf. (£27.95, US\$47.95.) Cambridge: Cambridge University Press on behalf of the MacKeith Press. 1997. ISBN 1-898-68313-1.

Neurofibromatosis type 1 (NF1) is one of the most common single gene disorders and it presents many difficult management problems. Physicians who have to tackle these problems in children will be stimulated and helped by this book, which presents the experience of a dedicated, multidisciplinary, paediatric NF1 clinic. The book starts with a brief historical overview and a highly readable summary of what is known of the molecular pathology of NF1. However, the core of the text is in a series of research paper style chapters which present data on a series of 200 children seen at the clinic. These chapters include a very useful review of the clinical features found in the patient group, a study on cognitive function and academic performance in affected children, a description of the MRI findings in a relatively unbiased group of the children, and a review of the perennially difficult problem of optic tract gliomas. A particularly intriguing chapter describes a significant (and controversial) association between MRI "unidentified bright objects" and cognitive impairment. Exploration of this area might provide some insight into the basic pathological mechanism of cognitive impairment in NF1. Last but not least, appendices contain a very useful clinical assessment protocol, and a very well written patient information sheet. Any doctor who is involved in the management of children with NF1 will find this book useful.

EVAN REID

Protein Targeting. Editor S M Hurtley. (Pp 214; £29.95.) UK: IRL Press. 1997. ISBN 0-19-963561-7.

In order fully to use the advantages subcellular compartmentalisation confers to an organism, the extensive obstacle of transporting proteins across lipid bilayers needs to be overcome. Protein targeting is the study of how newly synthesised proteins reach their designated cellular location for functionality and of the molecular conveyances used to do this. Targeting signals act as recognition motifs to permit the protein to bind to the

protein targeting machinery on the relevant organelle. Parallel themes are echoed throughout protein targeting pathways, both at the signalling and at the molecular apparatus level. For instance, targeting signals in proteins which arise directly from primary structure, are either partly or totally responsible for directing proteins to the endoplasmic reticulum, nucleus, peroxisome, and mitochondrion. Tertiary structural motifs also exist, such as the tyrosine containing b-turns which are specific for the targeting plasma membrane proteins to coated vesicles. Once at the target organelle, protein recognition by the appropriate transport machinery for internalisation is necessary. This may occur by endocytosis (at the plasma membrane) or through some sort of "pore" (for example, nucleus and endoplasmic reticulum). To ensure vectorial and irreversible transport, targeting signals may be destroyed at the final location of the protein. Of course the situation is far more complicated if the protein has to traverse several different intracellular membranes before arriving at its final location, each requiring a distinct signal. Lastly, we should not forget the vast support network of interacting proteins, such as molecular chaperones, which ensure correct folding and assembly of the newly targeted proteins. The understanding of protein targeting has many clinical applications, including the study of diseases which arise because of a fault in this mechanism and for elucidating appropriate intracellular pathways for targeted drug delivery for treating such diseases as colonic cancer.

This book is the 16th in the "Frontiers in Molecular Biology" series, which was devised to report on swiftly changing upfront areas of research in molecular biology. This volume certainly does this and it neatly complements two books on the same subject published by IRL Press in their "In Focus" (for students) and "Practical Approach" (with experimental protocols) series. I feel privileged now to own all three. "Protein Targeting" is an eminently readable book, by being brief, but maintaining attention to detail and by incorporating all the salient features of protein targeting mentioned above. It is as up to date as it can be for such a dynamic discipline with individual chapters written by distinguished scientists in the field. Protein targeting to all the major organelles is covered, resulting in a comprehensive volume informing the reader of everything they should ever want to know about eukaryotic protein targeting, but didn't know who or where to consult. As the editor explains in her preface, the less taxonomic trendy phyla have been omitted, apparently for space reasons. This confession of neglect is non-deleterious to the quality of the book

which holds together well as a whole, and the chapters can be read either as discrete entities or by cross referencing using the well thought out contents and index pages. The conciseness of this volume dictates that it is only suitable for the experienced cell biologist who has already committed to memory the acronyms rife in today's cell biology, but who lacks details beyond their exact subject.

Presentation throughout the book is generally very good. The format is easy to read; large font text is always favourable for those of us who have peered too long down our microscopes. Each chapter is well organised, systematically introducing the chosen targeting pathway before delving into the nitty-gritty and each ending with a comprehensive reference section. I thought the figures a little over simplified for the directed readership, who are generally over imaginative. However, the tables listing identified amino acid targeting signals provide easy access for the impatient scientist wanting to compare a newly identified signal sequence.

My major criticism of the book is the lack of any universal attempt to link the knowledge gained on studying eukaryotic protein targeting pathways to known clinical disorders. This is particularly infuriating in light of the fact that the study of some of these diseases was instrumental in elucidating molecular mechanisms of protein targeting pathways. A universal absence of protein targeting is incompatible with organelle biogenesis and therefore life, but defects in specific targeting signals which lead to disease are well characterised. For instance, in mucopolysaccharidosis II, newly synthesised lysosomal enzymes are mistargeted to the external environment; in familial hypercholesterolaemia, mutations in the low density lipoprotein (LDL) receptor reduce its endocytic ability, ultimately raising plasma LDL levels, and the commonest mutation reported in the cystic fibrosis transmembrane conductance regulator prevents it from leaving the endoplasmic reticulum for the cell surface, and therefore cystic fibrosis arises not so much from what the mutant protein is unable to do, but from the fact that it is now mistargeted. A short paragraph in each chapter updating the reader on the progress made on the medical relevance of their findings would have been welcome.

Overall, I recommend this book wholeheartedly and I am sure it will sit on the bookshelves of many cell biologists. It is an unusual scientific text in that you can actually read it rather than refer to it and it will certainly stimulate the desire for further reading.

JULIET A ELLIS