institutes. I doubt that it will be required reading for those who only seek to use AFP in the prevention of neural tube defects or in refined screening for Down’s syndrome.

D J H Brock

Recent Advances in Inborn Errors of Metabolism

This book is the proceedings of the Fourth International Congress of Inborn Errors of Metabolism, which was held in Japan in May 1987. Books like this can be out of date by the time they appear, containing nothing new. However, the editors are to be commended for ensuring that the papers have been published as a printed volume so quickly. A wide range of highly topical subjects has been selected and all the chapters written by those who are active in research, providing an authoritative view of their subject. There are sections on recent progress in the diagnosis and treatment of organic acidaemias, urea cycle disorders, lysosomal storage disorders, and various aspects of phenylketonuria, including the future for gene therapy. Several chapters are devoted to the molecular basis of these disorders and to new mechanisms of inborn errors including peroxisomal disorders and defects of processing of enzymes after polypeptide synthesis (post-translational disorders). There are also review chapters describing the identification of the cystic fibrosis gene locus and on the clinical and metabolic aspects of non-ketotic hyperglycaemia. The chapters are generally short, pithy, and of a high standard.

This is an excellent volume that achieves its goal of informing about recent advances. I strongly recommend it to all those looking for an update in inborn errors of metabolism.

J V Leonard

Genetics and Epithelial Cell Dysfunction in Cystic Fibrosis

For the past five years basic research on cystic fibrosis has been dominated by two themes. One has concentrated on the most notable expressed abnormality in the disease, the disturbance of chloride transport in epithelial cells. By dissecting and analysing aberrant chloride uptake in ever greater depth, it has been hoped to discover the specific protein abnormality which underpins the plethora of signs and symptoms that characterises cystic fibrosis. The other approach, called reverse genetics, seeks to move from linked DNA markers to the mutant gene itself, and then by cloning, sequencing, and data base comparisons, to make deductions about the nature and function of the expressed protein product.

Both approaches have attracted powerful groups of investigators. Although many of the currently solved inborn errors of metabolism have yielded to protein analysis, more intractable disorders like muscular dystrophy needed reverse genetics. In chronic granulomatous disease there was a splendid and triumphant fusion of the two approaches. In cystic fibrosis it is entirely probable that the validation of any candidate gene may need a substantial input from transport physiologists.

This book, the proceedings of a symposium held in Canada in November 1986, is primarily concerned with physiological investigations of chloride transport in epithelial cells. Nine of the 17 chapters record the various investigations being made on sweat glands, and on respiratory and tracheal epithelium. Aficionados of the work of Quinton, Boucher, Riordan, Pedersen, Frizzell, and Widdicombe will find little that has not been published elsewhere, but may enjoy having it all reviewed in a single text. Michael Welsh’s laboratory is not represented, a curious omission.

Molecular genetics is covered in three chapters. This subject is moving so fast that any conference proceedings are likely to be little more than an historical footnote to the search for the cystic fibrosis gene. The contribution from Bob Williamson’s group deals mainly with strategies, and disappointingly predates the discovery of closely linked markers which are already in routine use in most service laboratories. No mention is made of the first candidate gene which so excited the scientific world when it was published in Nature in 1987.

It is reasonable to expect that the costs of important conferences should be partly defrayed by the publication of their proceedings. Those not able to attend (this one was by invitation) can savour the atmosphere and catch up with the state of the art in a fast changing field. However, given the straitened financial circumstances of most university and hospital libraries, conference organisers have a heavy responsibility to publish timely and in full. These proceedings carry no discussions of the submitted papers and have taken almost a year to appear. I think it would be unreasonable to recommend the book to any but the most committed and involved cystic fibrosis research groups.

D J H Brock