The possibility that the loss of mutant alleles is balanced by the transition of larger normal ones into the lower DM range, which are more frequently found in DM males, should also be considered. Although our data are at variance with the recent report of Chakraborty et al. for normal subjects, it is still unknown whether the mechanisms responsible for segregation distortion are the same for DM alleles in the normal range compared to full scale expanded ones. The preferential transmission of larger alleles could be modulated by as yet unidentified cis and trans acting genetic elements, or could be a direct consequence of the CTG repeat number, affecting gamete or zygote viability. The recent observations that the expanded allele is preferentially transmitted to male meiosis in other conditions caused by dynamic expansions, such as dentatorubral-pallidolysian atrophy and MJD, 19,21 are more suggestive of a direct consequence of the CTG repeat number in these conditions, at least for full scale expanded alleles.

In summary, results from the present study confirm that the issue of meiotic drive and segregation distortion is still controversial in genetic counseling and may vary in different populations. Further studies will be important to shed more light on this fascinating question.

We are very grateful to Dr Rita C, M Pawlanello and Dr Suely K Marie for clinical and neurological examinations, to Marta Canovas and Constanza Urbana for their invaluable help, and to Dr Paulo A Otto for help in the statistical analysis. The support of FAPESP, CNPQ, FINEP, PRONEX, and IEAE is gratefully acknowledged.

BOOK REVIEWS

If you wish to order or require further information regarding the titles reviewed here, please write to or telephone the BMJ Bookshop, PO Box 295, London WC1H 9JR. Tel 0171 383 6244. Fax 0171 383 6662. Books are supplied post free in the UK and for BEPO addresses. Overseas customers should add 15% for postage and packing. Payment can be made by cheque in sterling drawn on a UK bank or by credit card (Mastercard, Visa, or American Express) stating card number, expiry date, and full name. (The price and availability are occasionally subject to revision by the Publishers.)


I have never been keen on problem based learning and I was quite sceptical when I started to read this book. The book covers most aspects of clinical genetics in 10 chapters. Each of these deals with a different aspect of genetics, such as X linked genetic transmission or mitochondrial inheritance, and is based around a clinical scenario. The book really deals with the clinical genetic transmission, for example, through the features of Duchenne and Becker muscular dystrophy (DMD), dystrophy, and X chromosome inactivation by discussing a couple who attend a genetic clinic for counselling because the woman’s half brother has DMD. This clinical scenario is taken in stages and the scientific basis for each aspect discussed. There is also a small section written by the mother of a child with DMD explaining what it is like to live with a child with DMD.

The book is very well written, very readable, and the numerous diagrams simple and explanatory. The conditions discussed in each chapter have been well chosen and the sections written by patients are excellent. After reading the book I have warmed to this method of learning. The facts are more memorable because the clinical applications are clear and because they all relate back to the couple or person discussed. I am not sure who would benefit most from this book. It is probably too clinical for undergraduates but I would recommend it to clinical students or clinicians wanting to learn more about genetics in practice.

DOROTHY TRUMP


This book describes molecular processes associated with dementia and is directed at scientists interested in the field. The molecular slant will decrease its appeal for most geriatric psychiatrists and geriatricians.

The 18 chapters consider a wide variety of processes which are involved in the pathology of Alzheimer’s disease and other dementias, ranging from genes involved in early onset familial Alzheimer’s disease and a review of possible biologic agents, to the consideration of β-amyloid metabolism, tau, energy metabolism, apoptosis, free radicals, inflammatory mediators, zinc metabolism, and transgenic approaches. There are also chapters on vascular dementia, Pick disease, and prion diseases. The editors and the authors are well respected workers in the field.

After reading the book, I felt slightly overwhelmed by the many possible mechanisms which can account for aspects of Alzheimer’s disease pathology, which are supported to various degrees by experimental data. I found the writing of the book to be clear and well written and was impressed by the well balanced arguments and lack of dogma. In general, the chapters are well written and scientifically detailed. Referencing is comprehensive. Appropriate diagrams and tables are provided.

One of the possible hazards of producing a book in a field which is witnessing rapid progress is that it may become out of date. This book is impressively up to date and many of the authors have been involved with research which was unpublished at the time of going to press. I will continue to find the book useful for a number of years because of its breadth and the chapters which concentrate on areas I am less familiar with. I recommend this book to those interested in the molecular mechanisms of dementia— I will be reading many sections again.

DAVID C RUBINSZTEIN


This collection of five papers comprises the proceedings of a day long seminar held in Western Australia in 1994, ostensibly for non-scientists. It describes the latest developments in reproductive technology together with the genetic techniques which are being applied to the field, with the aim of enabling the community at large to engage in the debate about the social and ethical implications. We have been grappling with the ramifications of mid-trimester prenatal diagnosis for many years; now there is the immediate possibility of preimplantation genetic testing and selection which raise additional issues. Processes which may be involved in making these decisions carry minimal risks, but they occupy less than 30 pages. The first is a sound “no nonsense” explanation of exactly what scientists are trying to achieve by these techniques and what they are not attempting to do. It is a pity the author of the last paper
