tion to the clinical presentation and pathology of A-T. Four sections follow: the first deals with the DNA defect in A-T whether regarded as a repair defect or a defect in replication synthesis; secondly, a section on genetics and cytogenetics; thirdly, several contributions on neuropathology; and finally a section on A-T and the immune system. Each of the chapters presented to the meeting has a discussion which I found quite helpful.

In the first section, on the possible DNA repair defect in A-T, the authors concluded disappointingly that this remains unproven. The balance of evidence does, however, seem to suggest that these cells are defective in some form of repair. Another important consideration for the participants was whether or not heterozygotes could be detected by increased cellular radiosensitivity. The conclusion was that this was not possible, but that it would soon be possible. In the two years or so that have passed since then, there is still no good evidence that these carriers can be adequately detected. The genetics of A-T receives attention in the book, but perhaps not as much as it deserves, especially after the statement by Swift that the consanguinity rate among his A-T families was much lower than they had found in similar studies on xeroderma pigmentosum or Fanconi's anaemia families. No further details are given. The recessive nature of the gene was not questioned. There is some discussion of gene frequency with respect to genetic complementation groups. The cytogenetic section is sparse and not very satisfactory. Two chapters have been added to update the observation (since 1984) that the chromosomal breakpoints in A-T translocations are at the sites of immune system genes. One inserted chapter contains the observation of a t(2;14) (p12;q32) translocation cell in PHA stimulated A-T lymphocytes without further comment.

Curiously I found this book stimulating except for one or two items it omitted to expand on. Those interested in A-T will be familiar with the content of the book. Others should perhaps try to borrow the book.

A M R TAYLOR

Advances in Human Genetics

Volume 15. Edited by H Harris and K Hirschhorn. (Figures + tables. £45.00.) New York: Plenum Press. 1986.

Four of the chapters in this book are of the type one has grown to expect from this series, comprehensive and authoritative reviews of an area of current active research. Two are of immediate clinical interest, one on chromosome abnormalities in

lymphomas and leukaemias by Le Beau and Rowley, and one on argininosuccinate synthetase and citrullinaemia by Beaudet and O'Brien and colleagues in Texas. The former offers a detailed but readable account of the evidence for consistent chromosome abnormalities in different leukaemias and lymphomas. It includes a brief discussion of the biological implications of these changes and also describes specific changes seen in leukaemia occurring in populations exposed to radiotherapy or environmental mutagens. The chapter on citrullinaemia covers a wide range of topics, from the differential diagnosis and clinical management of the patients to the analysis of heterogeneity in the mutations involved by determining the size of the mRNA transcripts.

A chapter on the molecular genetics of the HLA system, by Auffrey and Strominger, is a clear account of a confusing subject, providing the background knowledge required to appreciate new discoveries about function and relationship to disease. The fourth of these chapters, that on alcohol and aldehyde dehydrogenase by Moyra Smith, also describes an area where recent cloning of the gene promises exciting new developments in the understanding of gene expression and perhaps individual differences in response to alcohol.

The remaining one-third of the book is somewhat different, being largely a description of how Skolnick and Neel have been teaching their computer to recognise patterns of proteins after two dimensional gel electrophoresis. The objective is to produce a more sensitive and reliable indicator of human mutation rates than anything currently available. Any member of that small group of workers who have looked for mutations by screening samples on gels will appreciate that observer boredom is an obstacle well worth overcoming. It is a challenging chapter which should provide a basis for wide ranging argument, both technical and philosophical. Experimental validation of the approach in mouse experiments is proposed; it will be interesting to see the results.

All human geneticists, whether based in the clinic or in the laboratory, would find this a valuable book, both as a worthwhile read and a useful source of references.

S POVEY

Human Hemoglobin Genetics

By George R Honig and Julius G Adams III. (Pp. 452; figures + tables.) Wien, New York: Springer-Verlag. 1986.

Long time devotees of haemoglobin, used to hearing

of its impending demise as a subject worthy of serious study, will not be surprised to learn of yet another book which, like many of its preceding genre, manages nicely to refute the gloomy prognostications.

Human Hemoglobin Genetics is a welcome addition to what is already a substantial library given over to this remarkable protein, one of modern biology's more enduring workhorses. In their preface the authors say they have "endeavored to develop a relatively concise but detailed account of the current state of understanding of the hemoglobin system and the genetic and biological factors that govern its expression". I think they have succeeded admirably, keeping a nice balance between old and new, so that newcomers in the field are at least aware of the relevant background, and with sufficient detail and breadth to appeal both to students and practitioners. Inevitably, there are deficiencies. Once a topic is broached, where to draw the line can be problematical and I occasionally regretted the restrictions implicit in the title which, while keeping the size down to manageable proportions, put quite a lot of interesting comparative information from other species beyond the book's scope. A few of the sections, particularly those concerned with the haemoglobinopathies, are really too short to do their subjects justice. But these are minor quibbles that do not seriously mar what is a comprehensive and up to date survey of a very extensive field. There is also a very good bibliography and a useful appendix giving a sort of biological CV on each of the known human globin mutations.

Although not a specialist monograph, my review copy disappeared from my office often enough to suggest that the authors have perhaps been too modest in their expectations of the book's appeal. For those venturing into haemoglobin research for the first time, whether their interests lie in oncogenesis or population genetics, there is a great deal of interest in this excellent book. Old hands can also profit from it while smugly awaiting the next obituary.

J B CLEGG

Bone Marrow Transplantation for Treatment of Lysosomal Storage Diseases

Edited by W Krivit and N W Paul. (Pp 218; figures + tables. £35.00.) New York: Alan R Liss. 1986.

The remarkable progress made in recent years into the biochemical and molecular mechanisms of inherited disease has not been paralleled by develop-

ments in treatment. It remains a sad fact that curative therapy does not exist for most genetic disorders. Any sound approach to treatment or cure is to be welcomed. Many attempts have been made over the last 15 years to replace missing or defective enzymes in the metabolic storage diseases. To document the history of these attempts is to compile a catalogue of disappointments. However, the advent of bone marrow transplantation (BMT) has justifiably raised genuine hope that effective therapy may be close at hand.

Thus, it is timely that this slim volume, No 22 (1) in the Birth Defects Original Article Series, should now appear, presenting the proceedings of a symposium held in May 1985, attended by representatives of the centres at the forefront of these techniques. The text contains 14 succinct and eminently readable papers, immaculately reproduced on glossy paper and well illustrated. Subjects covered include the mucopolysaccharidoses (MPS), metachromatic leucodystrophy (MLD), adrenoleucodystrophy (ALD), Gaucher's disease, Lesch-Nyhan disease, Pompe's disease, technical aspects of transplantation, and studies in animal models.

The message which emerges is one of very cautious optimism. Biochemical correction, as judged by enzyme activity in white cells or plasma or both, was noted for all of the disorders under scrutiny. In several there was also evidence for clinical improvement as judged by resolution of airway obstruction (MPS I), joint immobility (MPS II and VI), ventricular overload (MPS VI), and peripheral nerve activity (infantile MLD).

Unfortunately, however, there was only limited evidence of significant change in the skeleton or in CNS function. No radiographical improvement in bone was noted for MPS II (nine months post BMT) or MPS VI (40 months post BMT). One child with MPS I was found to have normal CSF glycoaminoglycans levels 18 months after BMT, and evidence for arrest of regression based on developmental assessment was presented for five MPS I patients by the Westminster group. The mental state of the five year old child with infantile MLD was much better one year post BMT than that of his untreated sib at the same age, although serial CT scans revealed deterioration. Encouraging progress was also noted in the child with late infantile MLD. Sadly, neurological deterioration continued in the boy with ALD who died 20 weeks after transplantation, and there was no change in neurological status or behaviour in the patient with Lesch-Nyhan syndrome. Nor was any beneficial effect demonstrated in the infant with Pompe's disease who died 20 weeks after transplant.

There is a clear indication in these reports that BMT has something to offer in the treatment of