Storage diseases

Enzyme Therapy in Lysosomal Storage Diseases. (Proceedings of the Workshop on Cell Biological and Enzymological Aspects of the Therapy of Lysosomal Storage Diseases, Leiden.) Edited by J. M. Tager, G. J. M. Hooghwinkel and W. Th. Daems. Pp. xi+308. (North Holland: Amsterdam and Oxford; American Elsevier: New York, 1974.) Dfl65; $25.00.

THE identification of the lysosomal enzymes which are lacking in various human, genetically-controlled storage diseases, transformed the approach to this subject. Now that there is a large measure of agreement on the classification and manifestations of these diseases hope is rising that something might be done to ameliorate the conditions of those who suffer from the consequent physical abnormalities, mental retardation, pain and death. Most of the storage defects can be manifested in individual cultured cells which have enzymes missing and in which substrates consequently accumulate. Several of the defects can be identified in cultures of amnion cells, so that prenatal diagnosis is now feasible, which allows the possibility of induced abortion of homozygous children in affected families.

In April 1974 a workshop on the possibilities of enzyme replacement therapy was held in Leiden, and the papers presented were collected for publication in this volume. Many are concerned with the purification and further characterisation of the relevant enzymes and isoenzymes. Others are concerned with *in vitro* complementation, when cultured cells with different defects are fused to form heterokaryons or, remarkably, simply pick up enzymes from each other to attain phenotypically normal characteristics. Since the defect is in the lysosomal system, it may seem relatively easy to restore the missing enzyme, but replacement therapy presents serious difficulties. Liposomes have been used to introduce hydrolases into cells, where they can break down lysosomal contents in model systems. These can protect enzymes from inactivation by antibodies in the circulating blood.

Various attempts have been made to cure deficiency diseases by allotransplantation. In Fabry's disease accumulation of trihexosyl ceramide is often associated with deterioration of kidney function and, according to Desnick, kidney transplantation results in a fall in plasma trihexoside levels and the alleviation of painful crises. Although the missing enzyme, α-galactosidase A, is present in the urine, it is not demonstrable in blood, and the substrate is thought to be filtered from the blood and degraded in the kidney.

Enzymes administered by parenteral injection do not reach the central nervous system, so that a major problem will be to alleviate the effects of storage diseases there. If, however, levels of substrates in the blood could be reduced, there could be some beneficial effect.

The book's usefulness is increased by its rapid publication, made possible because the photo-offset process was used. The standard of reproduction of line illustrations is excellent and even that of half-tone illustrations is acceptable. I suspect that this will soon become the standard way to publish meeting reports. **A. C. Allison**

Introducing physics

Solid State Physics. (The Manchester Physics Series.) By H. E. Hall. Pp. xviii +351. (Wiley: London and New York, September 1974.) $15.00; £7.50.

THIS fourth book in the Manchester Physics Series is intended for use in undergraduate courses. The author has divided the text into two parts. In the first five chapters he presents a self contained, but elementary, introduction to the basic concepts of solid state physics, which should also be suitable for chemistry students. The early sections include topics such as crystal geometry, lattice vibrations, transport in semiconductors, free electron theory and magnetism. The remainder of the book is aimed towards honours physicists and contains chapters on band theory, X-ray and neutron scattering, thermal conductivity and Fermi surfaces. As a result of this division the formal constructs of reciprocal space and Brillouin zones do not appear until chapter six. The first chapter also flouts convention since the author attempts to explain why solids take up their various lattice structures rather than simply listing the possibilities. Indeed, the book begins with a detailed account of the quantum mechanics of covalent and ionic binding and the author follows this general line of argument throughout. The last few chapters on superconductivity, magnetic ordering and disordered solids are concise, but nevertheless, instructive. The section on the Fermi surface is, however, somewhat spoiled by the unwarranted amount of space (for a book of this kind) which is given to Overhauser's work on possible instabilities of the electron gas.

Each chapter has a useful set of problems associated with it, and answers are provided. I can recommend this as a good, if somewhat unusual, introductory text book. For a full honours course, however, one would probably require a little more experimental and theoretical detail than is contained here. **R. Evans**