lysosomal storage diseases, particularly those lacking CNS involvement. There has been no evidence for neuronal uptake following BMT in animals with lysosomal storage disorders and the evidence in humans is weak. The pioneers of these innovative techniques are to be congratulated. The papers in this text illustrate their awareness of the need for critical, objective, long term evaluation of the patients under their care although fuller data concerning morbidity and risks of mortality would be welcomed.

This book brings together a valuable collection of papers on a very important subject. Its successor providing details of longer follow up is eagerly awaited. Everyone involved in the care of these children will find this volume fascinating.

I D Young


The Mainwaring-Hughes Award, donated by the late Mr W T Mainwaring-Hughes, sometime Mayor of the County Borough of Swansea, is used by the University College of Swansea to commission people resident in Swansea or its locality to write pamphlets dealing with contemporary topics in Wales. As one of the regions in the United Kingdom with the highest prevalence of spina bifida and its associated conditions (neural tube defects), these abnormalities are a major contemporary problem in Wales. In the 1960s, one in every 120 babies born had a neural tube defect. Nansi James, who has been assisting Professor K M Laurence of the University of Wales College of Medicine, was invited to write an essay on the research work in South Wales into the cause and prevention of spina bifida.

The essay has nine short sections dealing with topics such as the nature of spina bifida, its effect on both the child and the parents, and the prevention of spina bifida in those who have already had an affected child and also in the population as a whole, by screening antenatal patients with serum alpha fetoprotein. To my mind, the most interesting section is the last one-third of the essay which adumbrates the research leading to primary prevention in mothers 'at risk' using dietary management, multivitamin preparations, and folic acid. This section also considers the economic aspects of prevention and looks forward to future strategies for prevention if the Medical Research Council's clinical trial succeeds in showing that folic acid or multivitamins or both can effectively prevent the recurrence of spina bifida and related abnormalities.

The author obviously had difficulty in knowing to whom to aim the pamphlet. From the nature of the background of the award, clearly the object of the essay was the general public. However, the detailed tables found in appendix IV, outlining the various outcomes of the individual studies, are aimed at a scientific audience. In addition, it is difficult to see the purpose, either for the layman or the scientist, of reproducing verbatim the 1720 and 1744 publications from The Philosophical Transactions of the Royal Society describing spina bifida.

It may be mean to cavil at this well meaning essay but, all the same, it would be negligent for a reviewer not to point out some flaws. The essay as a whole is less evaluative of published reports than one would like, the writer usually choosing to cite relevant works without commentary. Indeed, little recognition is given to other groups, particularly in the United Kingdom, involved in the prevention of spina bifida by periconceptional vitamin supplementation. Flaws in the design and analysis of the South Wales studies are glossed over. However, one cannot but admire the writer's enormous zeal. It is a pamphlet which can be thoroughly recommended to the general public, but not to the specialist in the subject.

Norman C Nevin