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ANIMAL MODELS OF INHERITED METABOLIC DISEASES

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Donald F. Patterson
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ANIMAL MODELS OF INHERITED METABOLIC DISEASES

Proceedings of the International Symposium on Animal Models of Inherited Metabolic Disease Held in Bethesda, Maryland October 19–20, 1981

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Preface

In October 1981, veterinarians, physicians and scientists from several continents convened at the National Institutes of Health to discuss their most recent studies on animal models of human disease. This symposium—"Animal Models of Inherited Metabolic Disease"—was sponsored by the Registry of Comparative Pathology of the Armed Forces Institute of Pathology, and by the Universities Associated for Research and Education in Pathology, Inc. The chapters in this book are based on the papers presented at the symposium.

The symposium was supported in part by a grant (RR 00301) from the Division of Research Resources, National Institutes of Health. In addition, we acknowledge the generous support of the Charles River Breeding Laboratories, Inc. and Hazelton Research Animals, Inc.

We hope that the scientific information recorded in this volume will not only provide a resource for veterinarians and physicians, but will stimulate efforts to identify and characterize new animal models of human disease. It is anticipated that the study of animal models will provide further insights into the molecular pathology of their human counterparts, and based on this information, the design and evaluation of new therapeutic strategies. Such studies should ultimately lead to better health care and effective treatment for the many patients and families who suffer from these unfortunate "experiments of nature".

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INTRODUCTION AND ACKNOWLEDGEMENTS

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It is my privilege to introduce and welcome you to this symposium and to what we believe will be a very lively and thought provoking monograph. The monograph is designed to provide the biochemical scientific community with new information and innovative possibilities for developing new animal models of genetic disease.

The Scientific Advisory Committee of the Registry, which I chaired at that time, developed the concepts for this symposium. It was recognized that there was a great unfulfilled need for "recognition and preservation of genetic disorders which otherwise might be lost." It was the hope that many of the mutations which now go unrecognized would be observed and preserved if it became clearer to veterinarians and pathologists what some of the needs are. It also seemed to us at that time that the types of clues of linkage of genetic models to hair coat, odor, age at death, enzyme and electrophoretic parameters, etc., needed to be more widely recognized and utilized if astute observation and follow-up were to help in the identification and preservation of new and valuable mutations which might otherwise be inadvertently discarded.

This was planned as a companion for the "Workshop on Needs for New Animal Models of Human Disease" which was held here in 1980 under the leadership of Dr. Donald Hackel. The resulting publication in the American Journal of Pathology and widespread distribution, along with much of the excellent discussion that occurred, has been widely

recognized as an important contribution to comparative pathology and to the experimental pathology of human disease of the future.

The grant which supports the Registry of Comparative Pathology is from the Animal Resources Branch, Division of Research Resources, National Institutes of Health. I do want to acknowledge and highlight this support because it has been seminal in the many pioneering efforts that this Registry has successfully undertaken. Dr. Dante Scarpelli and I have been the principal links of this Registry to the Board of Directors of Universities Associated for Research and Education in Pathology which has, from the Registry's inception, been its main sponsor and has, over and above our grant-related relationship, recognized the value of comparative pathology and provided a constant source of intellectual support and interest which needs to be emphasized as we open this symposium. Also providing valuable support and use of its facilities is the Armed Forces Institute of Pathology where the Registry is located.

The committee that set about to develop this program included not only the Scientific Advisory Committee of the Registry, chaired by Dr. Dante Scarpelli and consisting of Drs. C.C. Capen, D.B. Hackel, T.C. Jones, R.M. Lewis, and G. Majno along with staff (Drs. G. Migaki and S.R. Jones), but also three valuable special consultants, Drs. D.F. Patterson, R.J. Desnick, and G.A. Hegreberg. They have developed a program which we feel provides an excellent reflection of the ultramodern contributions being made in the areas of genetic modeling for the understanding of human diseases. We believe that the contributors they have chosen and the time provided for free-flowing discussion should lead to new concepts and plans for recognizing and preserving badly needed genetic models of disease.

Finally, I want to recognize several hidden heroes and heroines of this planning process and of the valuable volume which it has yielded.

First and foremost, there is Dr. George Migaki, the Chief Pathologist for the Registry of Comparative Pathology. As is true with almost everything the Registry does, Dr. Migaki has played a key role in planning, implementing and preparing these proceedings for publication. In this he has been assisted, not only by the Scientific Advisory

Committee to the Registry and the special consultants mentioned above, but also by Merryanna Swartz, his able Research Assistant, and Charmaine Goetz, the most capable Senior Secretary of the Registry. We also want to acknowledge with gratitude the financial assistance of the Charles River Breeding Laboratories and Hazleton Research Animals, Inc.

We have had extraordinary help from Dr. John Holman of the Animal Resources Branch in developing the arrangements for this meeting in Building 31 at the National Institutes of Health. We appreciate the skillful work of John Bowers and his co-workers in recording and transcribing the discussions so these could be corrected for publication while the participants are in attendance.

A special dividend of this symposium and its publication will be an exemplary "Compendium of Inherited Metabolic Diseases in Animals," worked up by Dr. Migaki and his staff with special assistance from Charlotte Kenton of the National Library of Medicine. Included are many models and references gleaned from the brand new "Bibliography of Naturally Occurring Animal Models of Human Disease" (and its companion volume on Induced Animal Models) edited by Drs. Gerald Hegreberg and Charles Leathers, Washington State University School of Veterinary Medicine in Pullman. It was also valuable, as Dr. Migaki points out in his introduction to the compendium and its bibliography, to be able to utilize the rather recent two volume work on "Spontaneous Animal Models of Human Disease" published by Academic Press and compiled by Drs. E.J. Andrews, B.C. Ward, and N.H. Altman. Of course, he utilized our favorite source, i.e., the Registry's own handbook: Animal Models of Human Disease, which has now developed into a valuable resource with ten fascicles and 232 models, many of which are of special relevance to this monograph.

Thus begins what Dr. Scarpelli and I believe will be a most successful symposium which, if we all do our part, should bring together the best thinking of ways to increase the yield of Animal Models of Inherited Metabolic Diseases which will be identified, preserved and utilized in the next ten years.

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