



Preface

Prion diseases



Bernardino Ghetti, MD



Pedro Piccardo, MD

Guest Editors

Progress in the field of prion disease has occurred at a staggering pace. One of the first keys that led to the concept of transmissible spongiform encephalopathies and the formulation of the prion hypothesis was the discovery of neuropathologic similarities between Creutzfeldt-Jakob disease, kuru, and scrapie. The prion hypothesis was formulated in the early 1980s; since then we have acquired a vast amount of knowledge of the prion protein and the various forms of related diseases (eg, idiopathic, genetically determined, and infectious). As the concept of the role of the prion protein in the pathogenesis of these diseases has evolved, new laboratory technologies have been developed that allow for an even greater understanding of its structure and function. During the past two decades, human and animal prion diseases have emerged that will present challenges well into the foreseeable future. At the same time, the development of therapeutic agents for some prion diseases is on the horizon.

Prion diseases pose a serious threat to human and animal species. The discovery in the United Kingdom of an epidemic of bovine spongiform encephalopathy (“mad cow disease”) and the detection of infected animals throughout Europe, Japan, and Israel has brought this issue to the public’s eye. It has been estimated that during the 1980s and 1990s, approximately 2 million cattle infected with bovine spongiform encephalopathy may have entered the human food chain. Compelling evidence has been obtained during the last several years to suggest that bovine spongiform encephalopathy has been passed to humans as the variant Creutzfeldt-Jakob disease, which was first described in the United Kingdom in the mid-1990s.

This issue of the *Clinics in Laboratory Medicine* features 12 articles written by leading scientists. Its purpose is to present pathologists with an overview of the current status of knowledge related to the biology of the prion protein and diseases associated with the presence of its misfolded form. A large portion of the issue focuses on the human prion diseases, including: clinical, pathologic, biologic, and molecular genetic characteristics; a description of most of the diagnostic methodologies available today; discussion on the safety of blood products; and the emergence of therapeutic strategies. The remaining articles deal with scrapie, chronic wasting disease, animal models of human prion diseases, the structure of the prion protein, and the conversion of the prion protein to the pathologic form.

Our hope is that this issue of the *Clinics in Laboratory Medicine* will in some way help pathologists face the coming challenges presented by these diseases.

Bernardino Ghetti, MD

Pedro Piccardo, MD

Department of Pathology and Laboratory Medicine

Division of Neuropathology

Indiana University School of Medicine

635 Barnhill Drive, MS A142

Indianapolis, IN 46202, USA