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| 1. Record Nr. | UNINA9910960105003321 |
| Titolo | Advancing prion science : guidance for the National Prion Research Program, interim report / / Rick Erdmann and Laura Sivitz, editors ; Committee on Transmissible Spongiform Encephalopathies: Assessment of Relevant Science, Medical Follow-Up Agency, Institute of Medicine of the National Academies |
| Pubbl/distr/stampa | Washington, D.C., : The National Academies Press, 2003 |
| ISBN | 9780309168595 0309168597 9780309504409 0309504406 |
| Edizione | [1st ed.] |
| Descrizione fisica | xx, 105 p |
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| Disciplina | 616.8/3 |
| Soggetti | Prion diseases - Research - United States Prion diseases - Diagnosis - United States Medicine, Military Public health - United States Slow virus diseases - Prevention Creutzfeldt-Jakob disease - Prevention Bovine spongiform encephalopathy Prion Diseases - prevention & control Public Health Prion Diseases - diagnosis Military Medicine Public Policy Health Policy United States |
| Lingua di pubblicazione | Inglese |
| Formato | Materiale a stampa |
| Livello bibliografico | Monografia |
| Note generali | Bibliographic Level Mode of Issuance: Monograph |
| Nota di bibliografia | Includes bibliographical references. |
| Nota di contenuto | Front Matter -- Reviewers -- Preface -- Acknowledgments -- Contents |

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Sommario/riassunto

In Advancing Prion Science, the Institute of Medicine's Committee on Transmissible Spongiform Encephalopathies Assessment of Relevant Science recommends priorities for research and investment to the Department of Defense's National Prion Research Program (NPRP). Transmissible spongiform encephalopathies (TSEs), also called prion diseases, are invariably fatal neurodegenerative infectious diseases that include bovine spongiform encephalopathy (commonly called mad cow disease), chronic wasting disease, scrapie, and Creutzfeldt-Jakob disease. To develop antemortem diagnostics or therapies for TSEs, the committee concludes that NPRP should invest in basic research specifically to elucidate the structural features of prions, the molecular mechanisms of prion replication, the mechanisms of TSE pathogenesis, and the physiological function of prions' normal cellular isoform. Advancing Prion Science provides the first comprehensive reference on present knowledge about all aspects of TSEs' from basic science to the U.S. research infrastructure, from diagnostics to surveillance, and from prevention to treatment. This report summarizes the progress thus far.