

Index

A

Aberrant folding, 103
Alzheimer's disease, 17, 24, 101, 112, 153, 240, 286
Amplification reactions, 121
Amyloid fibrils, conformational polymorphism, 153
Amyloidogenic region, 59
Amyloids, 147, 216, 265
 non-heritable, 273
Amyloid seeding assay (ASA), 121, 127
Anolis caroliensis, 230
Anxiety, 107
Atypical/Nor98 scrapie, 23

B

Biophysical monitors, 199
Bovine amyloidotic spongiform encephalopathy (BASE), 27, 37, 84
Bovine spongiform encephalopathy (BSE), 1, 23, 24, 51, 102
 atypical, 36, 84
BSE-C, 27, 37
BSE-H, 23, 38
BSE-L, 23, 37

C

C-terminal domain, 140
Candida albicans, 267
Cannibalism, 23
Carnivores, transmission, 68

Caspases, 260
Cattle, 13, 24, 36, 102, 170, 233, 238
 prion strain diversity, 84
Cell-free conversion, 121, 122
Cervid *prnp* gene, 59
Cervids, genetic susceptibility, 62
 prions, 51
 horizontal transmission, 66
 strain diversity, 85
Chemical chaperone, 187
Chemokines, 16
Chronic wasting disease (CWD), 26, 51, 102, 226
 diagnostics, 70
 strains, 69
 transmission, 85
Clusterin, 17
Conditional protein splicing, 207
Copper binding, 142, 199, 215
Creutzfeldt–Jakob disease (CJD), 1, 102, 225
 iatrogenic (iCJD), 23
 sporadic (sCJD), 4, 23, 84, 88, 127, 227
 variant (vCJD), 23, 27, 51, 62, 103
Cu/Zn superoxide dismutase, 107
Cysteine protease, 260
Cytokines, 16
cytoPrP, 105

D

D178N, 3, 5, 182
Deer, 52, 53, 68

Deer mice (*Peromyscus maniculatus*), 66
 Differential expression, 16
 Disease-related mutations, 169
 Doppel, 109, 225, 227
 Dorsal motoric nucleus of the vagus nerve (DMNV), 33, 57
 Drowsy (DY), 25, 152

E

E200K, 3, 6
 Elk, 52, 62, 85
 Embryogenesis, 250
 Encephalopathies, 121
 Environmental reservoirs, 58
 Environmental triggers, 285
 ER-associated degradation (ERAD), 105
 Erythroid differentiation-related factor (EDRF), 16
 European red deer (*Cervus elaphus elaphus*), 53, 64, 68
 Excitotoxins, 108
 Exotic ungulate encephalopathy (EUE), 102
 Expressed protein ligation (EPL), 199, 204
 E3 ubiquitin ligase HECTD2, 1, 15

F

Fatal familial insomnia (FFI), 2, 5, 29, 102, 182
 Feline spongiform encephalopathy (FSE), 102
 Fission yeast, 267
 Folding, 103, 135, 143
 Fungal prions, 257

G

Gain-of-function, 249
 Gametogenesis, 250
 Genetic control, 286
 Genetic susceptibility, 6
 Genome wide association studies (GWAS), 7
 Gerstmann–Sträussler–Scheinker syndrome (GSS), 2, 5, 28, 81, 102
 Glycoform profile, 25

Glycosylphosphatidyl–inositol (GPI) anchor, 59, 104, 110, 137, 170, 203, 232
 Gut-associated lymphoid tissue (GALT), 57

H

Hematoxylin–eosin (HE) staining, 57
 Histopathology, 56
 Hsp104, prion propagation, 279
 Human prion diseases, atypical, 28
 Human prions, 23, 171
 PRND, 239
 SPRN, 234
 strain diversity, 88
 Hyper (HY), 25, 59, 152

I

Immunohistochemistry (IHC), 31, 57, 70
 Incubation periods, 1, 9
 Inherited prion disease (IPD), 1, 3
 Interleukin–4, 16
 Intrinsically disordered, 101

K

Kaliotoxin, 203
 Kuru, 2, 7, 23, 30, 88, 102

L

Loss-of-function, 249

M

Macaques, 70
 Meadow voles (*Microtus pennsylvanicus*), 66
 Methionine129, 29
N,N'-(Methylenedi–4,1-phenylene)bis[2-(1-pyrrolidinyl)acetamide] (GN8), 188
 MicroRNAs (miRNAs), 18
 Mink prions, 25, 68, 85, 103
 hamster-adapted, 59
 Misfolding, 101, 103, 169, 179
 pH-induced, 169
 Molecular chaperone, 257

- Molecular dynamics simulation, 169, 172
 Monocyte chemoattractant protein-1 (Mcp1), 16
 Moose (*Alces alces shirasi*), 52, 66
 Mountain lions, 68
 Mouse (*Mus musculus*), 60, 66, 233
 models, 9, 78
 Sprn, 233
 Mule deer (*Odocoileus hemionus*), 52
Mus spretus, 233
- N**
 Native chemical ligation (NCL), 199, 200, 202
 Neurodegeneration, 101, 245
 Neuroprotection, 101, 108, 225
 Neurotoxic stressors, 108
 Nitrogen uptake regulation, 262
 Nor98, 32
- O**
 6-Octapeptide repeat insertion (OPRI), 3, 4
 Octapeptide repeat region, 138
- P**
 P102L, 3, 5
 Packing polymorphism, 156
 Pathogenic mutations, 182
 Periodic sharp wave complexes (PSWC), 29
 Peripheral chronic demyelinating polyneuropathy, 107
 Peroxiredoxins, 286
 PG14-PrP, 106
Pichia methanolica, 279
 Plasma membrane attachment, 242
Podospira anserina, 257, 259
 Point mutations, 5
 Polymorphisms, 1, 153
 Predators, 68
 Prion composition, 121
 Prion containment, 51
 Prion diseases, 23, 135
 inherited, 3
 Prion-forming domains (PrD), 260, 281
 Prion propagation, 257
 in vitro, 155
 Prion protein (PrP) 1ff, 169
 chemical synthesis, 207
 domain structure, 228
 interaction with small molecules, 199
 metals/peptides/small molecules, 214
 pH, 176
 Prion protein amyloid, structural models, 149
 Prion protein gene (PRNP), 1
 Prion proteins, conversion, 145
 folding, 135
 Prion replication, models, 145
 Prion seeded conversion, 121
 Prion strains, 23, 78, 135
 diversity, 84, 85
 Prion structural biology, 135
 Prions, 1
 environment, 58
 fungal, 257
 gene regulators, 261
 stability, 58
 synthetic mammalian, 155
 transcriptional regulation, 262
 types, 23
PRND gene, anatomical expression, 239
Prnp gene, 2, 10, 103, 226
PRNP gene, 2, 28, 227
 polymorphism, 82
 Propagons, 257
 Protease-sensitive prionopathy (PSPr), 31
 Proteinase K, 146
 Protein misfolding cyclic amplification (PMCA), 25, 58, 69, 71, 86, 121
 serial automated (saPMCA), 122
 Proteins, chemical synthesis, 199
 dynamics, 169
 folding, 135
 misfolding, 169, 179
 PrP^c, 2, 24, 59, 101, 121, 201
 physiological function, 107
 PrP^{Sc}, 2, 23, 58, 68, 121, 201
 biochemical/-physical characteristics, 146
 structural models, 149
 PrP^ΔHD, 106
 Prusiner, Stanley, 136
 Push-pull effects, 247

Q

- Quaking-induced conversion (QuIC), 121, 128
- Quantitative PMCA (qPMCA), 122
- Quantitative trait loci (QTLs), 12, 233
- Quasi-species, 79

R

- Racoons, 68
- Recombinant protein-misfolding cyclic amplification (rPMCA), 121
- Red-backed voles (*Myodes gapperi*), 66
- Red deer (*Cervus elaphus elaphus*), 53, 64, 68
- Regulators of cell function, 287
- Rocky Mountain elk (*Cervus elaphus nelsoni*), 52, 62
- Roe deer (*Capreolus capreolus*), 53
- rPrP amyloid fibrils, 150
- Ruminants, transmission, 67

S

- Saccharomyces cerevisiae*, 257
- Scavengers, 68
- Schizosaccharomyces pombe*, 267
- Scrapie, atypical/Nor98, 32, 83
 - murine, 106
 - sheep/goats, 82
 - transmission, 27, 67
- Scrapie-responsive gene (ScRG), 16
- Serial automated PMCA (saPMCA), 122
- Shadoo (shadow of prion protein), 8, 225
- Sheep (*Ovis aries*), 2, 23, 60, 67, 80, 123, 226
 - SPRN, 234
- Single nucleotide polymorphisms (SNPs), 7
- Small molecule ligands, 187
- Soil, 59
- Species barrier, 67, 68, 79, 85, 126, 268
- Sperm, 244
- Spongiform encephalopathies, 121
- Sporadic fatal insomnia (SFI), 88, 102
- Squirrel monkeys (*Saimiri sciureus*), 36, 70
- Stetsonville isolate, 25

Strains, 79

- Stress-protective protein, 108
- Susceptibility, 1
 - genetic, 6
- Synuclein, 104
- Syrian hamster (*Mesocricetus auratus*), 60

T

- Thioflavin T, 127, 147
- Transcriptional regulation, 262
- Transgenic models, 79
- Translational regulation, 263
- Transmissible mink encephalopathy (TME), 25, 87, 102
- Transmissible spongiform encephalopathies (TSEs), 24, 102, 121
- Transmission, 9, 27, 67, 81, 121, 259
 - natural, cervids, 66
- Trimethylamide N-oxide (TMAO), 187

U

- Ubiquitin-proteasome system (UPS), 286

V

- Validation, 18
- Variably protease-sensitive prionopathy (VPSPr), 31

W

- White-footed mice (*Peromyscus leucopus*), 66
- White-tailed deer (*Odocoileus virginianus*), 52
- Wildlife prion disease, 51

Y

- Yeast prions, 257

Z

- Zipper, 150, 155, 212, 275
- Zoonotic potential, 51, 69, 78