PRION DISEASE

Essay

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In Clinical & Chemical Pathology

By

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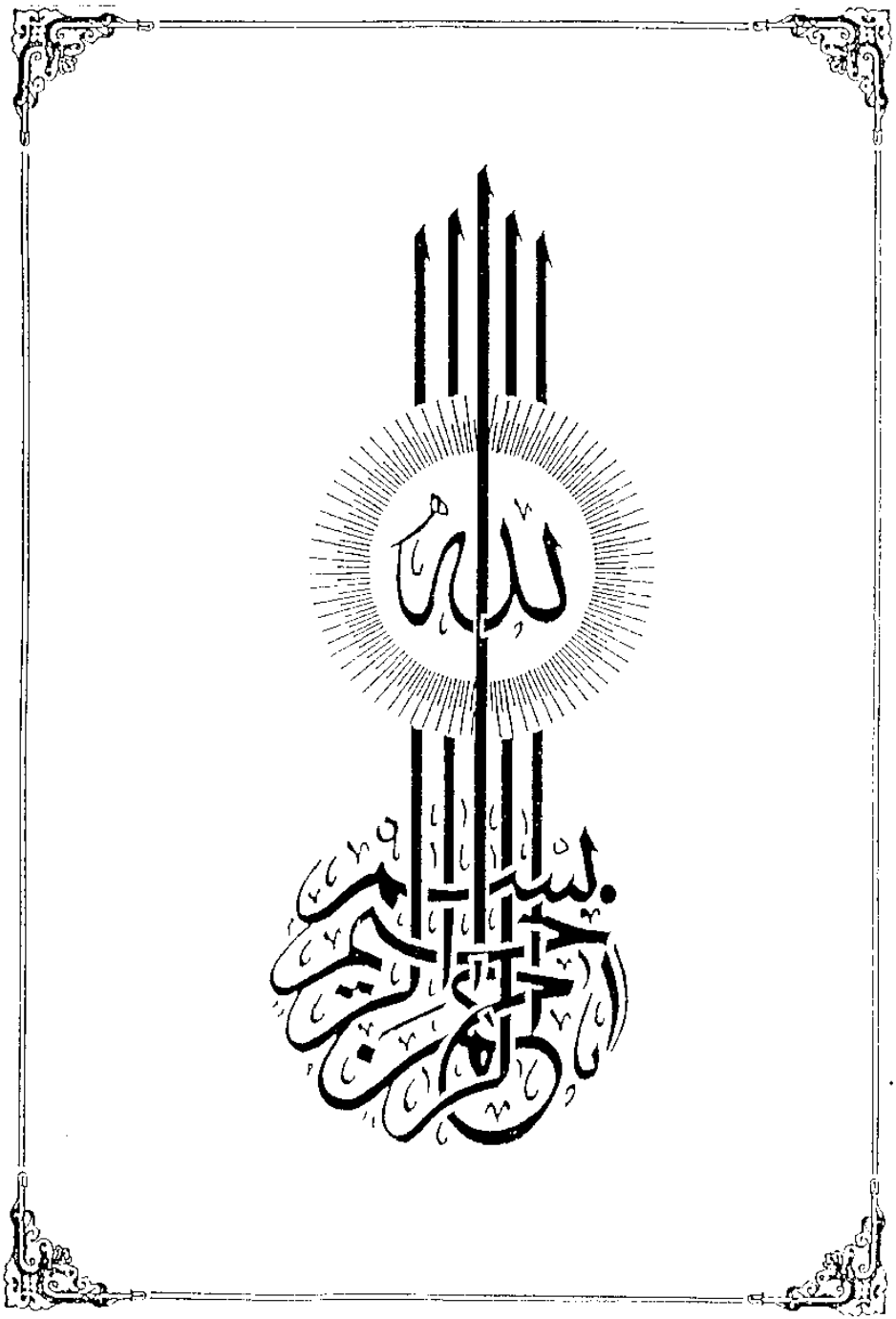

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List of Abbreviations

BSE	Bovine spongiform encephalopathy.
CJD	Creutzfeldt-Jakob disease.
CSF	Cerebrospinal fluid.
EEG	Electroencephalogram
EM	Electron microscope.
FFI	Fatal familial insomnia.
FSE	Feline spongiform encephalopathy.
GSS	Gerstmann-Straussler-Scheinker syndrome.
KD	Kilo dalton.
MSE	Mink spongiform encephalopathy.
nvCJD	new variant Creutzfeldt-Jakob disease.
PrP	Prion protein.
PrP^{c}	Prion protein cellular isoform.
PrP^{sc}	Prion protein scrapie form.
TSE	Transmissible spongiform encephalopathy.
UV	Ultra violet irradiation.
2-DE	2 Dimensional electrophoresis.

List Of Errata

P2	L3	its ----------→ the
P3	L2	an ----------→ are
P4	L9	disease ----------→ diseases
	L9	enter ----------→ enters
	L9	act----------→ acts
P5	L1	hypotheses --------→ hypothesis
	L3	resist ----------→ resists
	L15	adapts----------→ adopts
	L17	nucleus----------→ nucleo
P7	L11	were----------→ was
P9	L17	just----------→ first
P12	L16	progress----------→ progresses
P13	L1	cause ----------→ caused
P16	L7	accumulated -----→ accumulates
P17	L2	eve - ----------→ ewe
P21	L2	female ----------→ females
P23	L1	manifest ---------→ manifested
P32	L9	occur----------→ occurs
	L13	include ----------→ includes
P33	L11	causes----------→ cause
P36	L21	excepting--------→ accepting
P37	L19	respects----------→ aspects
P39	L18	patient----------→ patients
	L19	year----------→ years
P41	L1	types----------→ type
	L14	shows ----------→ show
P46	L9	a----------→ as
P49	L21	show ----------→ shows
P50	L2	make----------→ makes
P55	L4	pods----------→ rods
P56	L13	were----------→ was
	L15	markers---------→ marker
P60	L12	correlate-------→ correlates
P61	L8	in to ----------→ into
P63	L6	bindings-------→ binding
P66	L21	were----------→ are
P67	L16	deteriorated--→ deteriorates
P69	L9	depend -------→ depends
P74	L2	are----------→ is
P75	L2	follows-------→ either
P78	L7	in----------→ is

Contents

Introduction

Recent events have engendered great interest in Transmissible Spongiform Encephalopathies (TSE) or Prion disease. Several hypotheses have been advanced to explain the nature of the agent causing prion disease, including that it is a virino (which is a complex of protein and nucleic acid) or a small virus. However, prion (which is a protienaceous infectious agent without DNA and RNA) hypothesis has gained wide acceptance ***(Prusiner, 1994).***

All the prion diseases that have been for many years referred to as 'slow virus' or 'slow infection' share many features; as they are confined to the central nervous system, with prolonged incubation period ranging from several months to two or three decades. In each case, the disease inevitably progress to death ***(Gajdusek, 1985).***

Prions are the infectious agent for many diseases affecting a wide range of animal species sheep, goat (Scrapie), cattle (Bovine Spongiform Encephalopathy), and man (Kuru, Creutzfeldt-Jakob disease, Gerstmann-Straussler- Scheinker syndrome and Fatal Familial Insomnia) ***(Hart, 1996).***

Aim of the work:

The aim of this work is to review the subject of prion diseases with special emphasis on its laboratory diagnosis.