

Supporting Information

Berry et al. 10.1073/pnas.1317164110

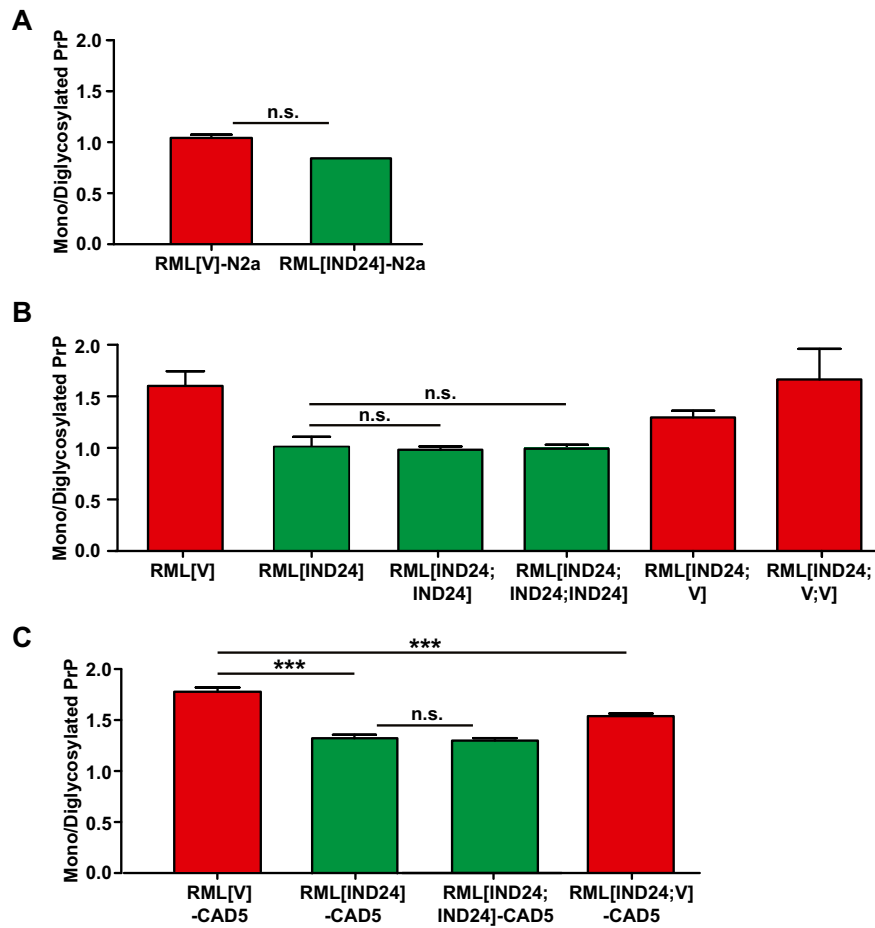


Fig. S1. Glycosylation ratios of prions from IND24-treated and vehicle-treated mice. Nomenclature as defined in the main text. (A) Average ratio of monoglycosylated to diglycosylated disease-causing prion protein (PrP^{Sc}) in N2a cells infected with RML[V] (red, $n = 2$) or RML[IND24] (green, $n = 1$). Error bar represents range. (B) Average ratio of monoglycosylated to diglycosylated PrP^{Sc} in RML[IND24]-infected mice treated with IND24 ($n = 7$); RML[IND24;IND24]-infected mice treated with IND24 ($n = 5$); RML[IND24]-infected mice treated with vehicle ($n = 7$); and RML[IND24;V]-infected mice treated with vehicle ($n = 6$), as indicated. The final treatment in mice is indicated as red (vehicle) or green (IND24). RML[IND24] and RML[V] from Fig. 1D are shown for comparison. Error bars represent standard deviation. n.s., not significant, Student's t test. (C) Average ratio of monoglycosylated to diglycosylated PrP^{Sc} in CAD5 cells infected with RML[IND24;V] ($n = 6$) or RML[IND24;IND24] ($n = 6$). The final treatment in mice is indicated as red (vehicle) or green (IND24). RML[IND24] and RML[V] from Fig. 2E are shown for comparison. Error bars represent standard deviation. *** $P < 0.001$; n.s., not significant, Student's t test.

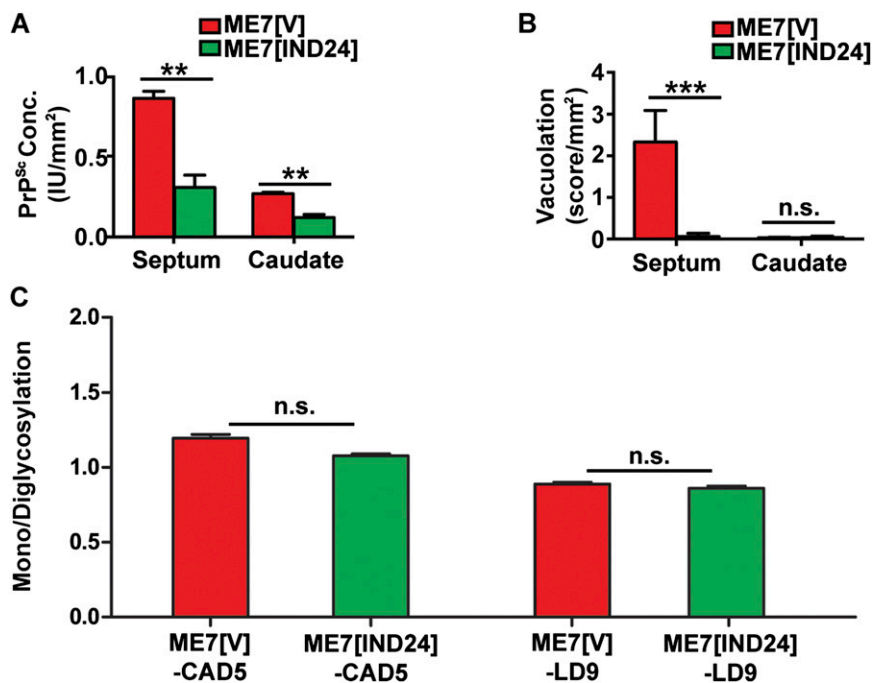


Fig. S2. Effects of IND24 treatment on ME7. Quantification of PrP^{Sc} deposition (A) and vacuolation (B) in the septum and caudate nucleus from ME7[V] (red; *n* = 5) and ME7[IND24] (green; *n* = 9) animals, as indicated. Error bars represent SEM. PrP^{Sc} measurements were made in intensity units (IU). ***P* < 0.01, ****P* < 0.001, n.s. = not significant, Student's *t* test with Bonferroni correction. (C) Average ratio of monoglycosylated to diglycosylated PrP^{Sc} in CAD5 and LD9 cells infected with ME7[V] (red; *n* = 3) or ME7[IND24] (green; *n* = 6). Error bars represent SD.

Table S1. Prion protein glycosylation ratios from passaged prions

Strain	<i>n</i>	Diglycosylated mean ± SD	Monoglycosylated mean ± SD	Unglycosylated mean ± SD	Mono-/diglycosylated ratio mean ± SD
RML[V]	7	0.31 ± 0.02	0.51 ± 0.02	0.18 ± 0.01	1.64 ± 0.14
RML[IND24]	7	0.36 ± 0.03	0.37 ± 0.01	0.27 ± 0.03	1.03 ± 0.09
RML[IND81]	9	0.43 ± 0.04	0.44 ± 0.03	0.14 ± 0.03	1.03 ± 0.17
RML[IND24;V]	7	0.33 ± 0.01	0.43 ± 0.01	0.24 ± 0.02	1.30 ± 0.06
RML[IND24;IND24]	7	0.37 ± 0.01	0.36 ± 0.01	0.27 ± 0.01	0.98 ± 0.03
RML[IND24;V;V]	6	0.29 ± 0.03	0.47 ± 0.03	0.24 ± 0.03	1.66 ± 0.12
RML[IND24;IND24; IND24]	5	0.36 ± 0.04	0.36 ± 0.02	0.27 ± 0.06	1.01 ± 0.09

Table S2. Prion infectability and IND24-resistance in cell culture

Inoculum	Cell line					
	N2a		CAD5		LD9	
	Infection*	Resistance to IND24 [†]	Infection*	Resistance to IND24 [†]	Infection*	Resistance to IND24 [†]
RML	3/3	+	2/2	+	—	—
RML[V]	2/3	+	6/6	+	—	—
RML[IND24]	1/8	+	6/6	++++	—	—
RML[IND24;V]	6/6	+	6/6	+	—	—
RML[IND24; IND24]	0/6	n/a	6/6	++++	—	—
ME7	—	—	5/5	+++	5/5	+++
ME7[V]	—	—	3/3	+++	3/3	+++
ME7[IND24]	—	—	6/6	+++	6/6	+++

n/a, not applicable; —, not attempted.

*Number of successful infections over the number of attempts.

[†]Qualitative measure of resistance, from low (+) to high (++++).