Correction: Cortical malformations are associated with a rare polymorphism of cellular prion protein

In the article “Cortical malformations are associated with a rare polymorphism of cellular prion protein” (*Neurology*® 2004;63:557–560), by R. Walz et al., the authors reported that a variant allele at the cellular prion protein gene (*PRNP*) Asn171Ser was associated with symptomatic epilepsies related to different forms of focal malformations of cortical development (MCD). Unfortunately, a second researcher in the group was unable to reproduce part of these results when evaluating 94% of the patient samples. The accurate data presented in table 1 demonstrate that the *PRNP* variant allele Asn171Ser is associated to MCD ($p = 0.005$), however with a lower prevalence than that previously described.

The Asn171Ser genotype was found in 7.7% of patients from Group I (abnormal neuronal and glial proliferation or apoptosis) ($p = 0.016$) and in 6.7% in those from Group III (abnormal cortical organization, including late neuronal migration) ($p = 0.021$) (table 2). A detailed description of the MCD subtypes and genotype at codon 171 is presented in table 3.

Therefore, the results show that although the polymorphism Asn171Ser is present in a lower frequency than previously described, its presence is associated with refractory epilepsy related to different MCD subtypes.

The authors deeply apologize for unintentionally permitting partially incorrect data to reach the scientific community.

Table 1 Frequency of *PRNP* alleles in patients with malformations of cortical development and controls

Residue position	Genotype	Exposed (%)		Crude		Adjusted	
		Controls, n (%)	Cases, n (%)	OR (95% CI)	p Value	OR (95% CI)	p Value
117	Ala/Ala	n = 174 168 (96.6)	n = 64 60 (93.8)	1.00		1.00	
	Ala/Ala _{silent}	6 (3.4)	4 (6.2)	1.87 (0.51–6.84)	0.346*	0.93 (0.17–4.96)	0.932†
129	Met/Met	n = 174 100 (57.5)	n = 64 27 (42.2)	1.00		1.00	
	Met/Val	66 (37.9)	27 (42.2)	1.51 (0.82–2.81)	0.187*	1.39 (0.72–2.67)	0.325§
	Val/Val	8 (4.6)	10 (15.6)	4.63 (1.67–12.87)	0.003*	3.03 (0.96–9.59)	0.060§
171	Asn/Asn	n = 174 174 (100.0)	n = 64 60 (93.8)	1.00	—	—	
	Asn/Ser	0 (0)	4 (6.2)	NA	0.005*	NA	NA
Octarepeat	R12234	n = 174 164 (94.3)	n = 60 [¶] 57 (95.0)	1.00		1.00	
	R12234/R1234	10 (5.7)	3 (5.0)	0.86 (0.23–3.25)	0.828*	0.86 (0.22–3.34)	0.827
	R1234	0 (0)	0 (0)	NA	NA	NA	NA

*Binary logistic regression.

†Fisher exact test.

‡Multiple logistic regression adjusted for gender, ethnicity, codon 129 distribution, and presence of the octarepeat deletion.

§Multiple logistic regression adjusted for gender, ethnicity, genotypes at position 117, and presence of octarepeat deletion.

^{||}Multiple logistic regression adjusted for gender, ethnicity, genotypes at position 117 and 129.

[¶]Only 60 samples could be evaluated for the analysis of the octarepeat.

NA = not applicable.

Table 2 Frequency of PRNP alleles in different types of MCD

Residue position	Genotype	Exposed (%)		Crude		Adjusted	
		Controls, n (%)	Cases, n (%)	OR (95% CI)	P Value	OR (95% CI)	P Value
Group I—Abnormal neuronal and glial proliferation or apoptosis (n = 26)							
		n = 174	n = 26				
117	Ala/Ala	168 (96.6)	24 (92.3)	1.00		1.00	
	Ala/Ala _{silent}	6 (3.4)	2 (7.7)	2.33 (0.44–12.23)	0.316*	1.13 (0.12–10.82)	0.918 [†]
		n = 174	n = 26				
129	Met/Met	100 (57.5)	9 (34.6)	1.00		1.00	
	Met/Val	66 (37.9)	13 (50.0)	2.19 (0.88–5.41)	0.090*	1.81 (0.71–4.64)	0.214 [§]
	Val/Val	8 (4.6)	4 (15.4)	5.55 (1.40–22.09)	0.015*	4.13 (0.91–18.68)	0.065 [§]
		n = 174	n = 26				
171	Asn/Asn	174 (100.0)	24 (92.3)	1.00		—	—
	Asn/Ser	0 (0)	2 (7.7)	NA	0.016*	NA	NA
		n = 174	n = 25**				
Octarepeat	R12234	164 (94.3)	25 (100.0)	1.00		1.00	
	R12234/R1234	10 (5.7)	0 (0)	NA	0.617 ⁺	NA	NA
	R1234	0 (0)	0 (0)	NA	NA	NA	NA
Group II—Abnormal neuronal migration (n = 8)							
		n = 174	n = 8				
117	Ala/Ala	168 (96.6)	8 (100.0)	1.00		1.00	
	Ala/Ala _{silent}	6 (3.4)	0 (0)	NA	1.00 ⁺	NA	NA
		n = 174	n = 8				
129	Met/Met	100 (57.5)	4 (50.0)	1.00		1.00	
	Met/Val	66 (37.9)	2 (25.0)	0.76 (0.13–4.25)	0.753*	0.40 (0.04–2.69)	0.420
	Val/Val	8 (4.6)	2 (25.0)	6.25 (0.99–39.50)	0.051*	5.10 (0.76–34.12)	0.093
		n = 174	n = 8				
171	Asn/Asn	174 (100.0)	8 (100.0)	1.00		1.00	—
	Asn/Ser	0 (0)	0 (0)	NA	NA	NA	NA
		n = 174	n = 8				
Octarepeat	R12234	164 (94.3)	8 (100.0)	1.00		1.00	
	R12234/R1234	10 (5.7)	0 (0)	NA	1.00 ⁺	NA	NA
	R1234	0 (0)	0 (0)	NA	NA	NA	NA
Group III—Abnormal cortical organization, including late neuronal migration (n = 30)							
		n = 174	n = 30				
117	Ala/Ala	168 (96.6)	28 (93.3)	1.00		1.00	
	Ala/Ala _{silent}	6 (3.4)	2 (6.7)	2.00 (0.38–10.41)	0.410*	1.03 (0.11–9.45)	0.981 [¶]
		n = 174	n = 30				
129	Met/Met	100 (57.5)	14 (46.7)	1.00		1.00	
	Met/Val	66 (37.9)	12 (40.0)	1.30 (0.57–2.98)	0.538*	1.27 (0.52–3.10)	0.605 [#]
	Val/Val	8 (4.6)	4 (13.3)	3.57 (0.95–13.42)	0.060*	1.98 (0.37–10.71)	0.430 [#]
		n = 174	n = 30				
171	Asn/Asn	174 (100.0)	28 (93.3)	1.00		1.00	—
	Asn/Ser	0(0)	2 (6.7)	NA	0.021 [†]	NA	NA

Table 2 Continued		Exposed (%)		Crude		Adjusted	
Residue position	Genotype	Controls, n (%)	Cases, n (%)	OR (95% CI)	p Value	OR (95% CI)	p Value
		n = 174	n = 27 [#]				
Octarepeat	R12234	164 (94.3)	24 (88.9)	1.00		1.00	
	R12234/R1234	10 (5.7)	3 (11.1)	2.05 (0.53-7.98)	0.301*	2.04 (0.50-8.29)	0.319**
	R1234	0 (0)	0 (0)	NA	NA	NA	NA

*Binary logistic regression.

[†]Fisher exact test.

[‡]Multiple logistic regression adjusted for gender, ethnicity, and genotype at codon 129.

[§]Multiple logistic regression adjusted for gender, ethnicity, and genotype at codon 117.

^{||}Multiple logistic regression adjusted for gender and ethnicity.

[¶]Multiple logistic regression adjusted for gender, ethnicity, genotype at codon 129, and presence of octarepeat.

[#]Multiple logistic regression adjusted for gender, ethnicity, genotype at codon 117, and presence of octarepeat.

^{**}Multiple logistic regression adjusted for gender, ethnicity, genotype at codon 117 and 129.

^{††}In Group I, only 25 samples could be evaluated for the analysis of the octarepeat.

^{†††}In Group III, only 27 samples could be evaluated for the analysis of the octarepeat.

NA = not applicable.

Table 3 Frequency of <i>PRNP</i> genotype at codon 171 among different forms of MCD			
Type of malformation of cortical development	All cases	Genotype at codon 171	
		Asn/Asn(%)	Asn/Ser(%)
I. Neuronal and glial proliferation or apoptosis (n = 26)			
A. Microcephalies (n = 0)			
1. Microcephaly with polymicrogyria/cortical dysplasia	None		
B. Abnormal proliferation (n = 26)			
1. Non-neoplastic			
a. Cortical hamartomas*	2	2 (100.0)	None
b. Cortical dysplasia with balloon cells	9	8 (88.9)	1 (11.1)
c. Hemimegalencephaly	5	4 (80.0)	1 (20.0)
2. Neoplastic (with cortical disordered)			
a. Dysembryoplastic neuroepithelial tumor	8	8 (100.0)	None
b. Ganglioglioma	1	1 (100.0)	None
c. Gangliocytoma	1	1 (100.0)	None
II. Neuronal migration (n = 8)			
A. Lissencephaly/subcortical band heterotopia spectrum (n = 6)	6	6 (100.0)	None
B. Heterotopia (n = 2)			
1. Subependymal (periventricular)	1	1 (100.0)	None
2. Subcortical (other than band heterotopia)	1	1 (100.0)	None
III. Cortical organization and late neuronal migration (n = 30)			
A. Polymicrogyria and schizencephaly (n = 18)			
1. Bilateral polymicrogyria syndromes	5	5 (100.0)	None
2. Schizencephaly (polymicrogyria with clefts)	3	2 (66.7)	1 (33.3)
3. Polymicrogyria with other brain malformations or abnormalities	10	9 (90.0)	1 (10.0)
B. Cortical dysplasia without balloon cells (n = 12)	12	12 (100.0)	None
Total	64	60 (93.8)	4 (6.2)

*Cortical hamartomas not related to tuberous sclerosis.

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