## Sporadic Creutzfeldt–Jakob disease: Clinical and diagnostic characteristics of the rare VV1 type

Meissner et al. report the clinical features of nine patients, all but one men, with the rare VV1 subtype of sporadic CJD. Patients presented with early onset slowly progressive dementia or behavioral abnormalities (median age 44 years, range 19 to 55). Diagnosis is supported by positive 14-3-3 protein in the CSF and cortical signal increase on MRI.

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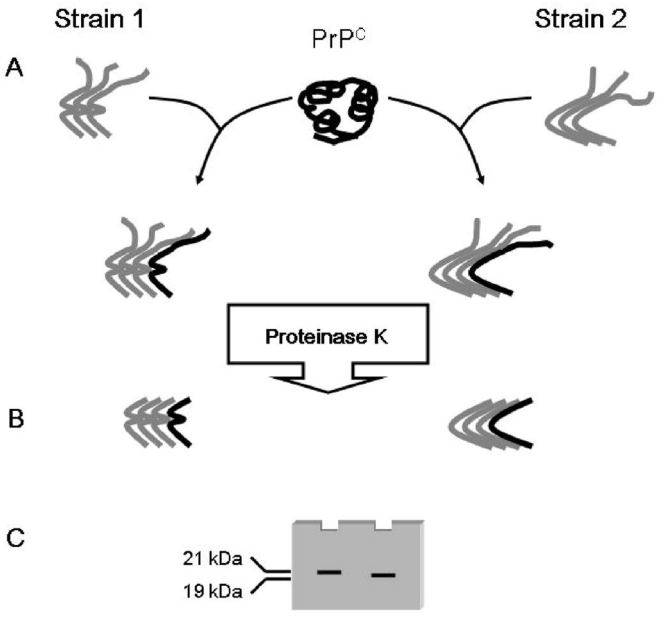


*The editorial by Patrick Bosque notes that in sporadic CJD, aggregated PrP ($PrP^{Sc}$) initially arises spontaneously, then interacts with normal PrP ($PrP^{C}$), causing it to misfold, recruiting it into the abnormal aggregate. The aggregate grows and then fractures, increasing the number of aggregates propagating throughout the CNS. Given this simple composition, the discovery of prion strains and CJD molecular subtypes poses a conundrum. Studies of prion strains have found that the size of the protease-resistant core differs slightly between some strains, suggesting that PrP can misfold and aggregate in different ways to create infectious prions. How this variable misfolding translates into the complex clinical and pathologic differences between strains is unknown, but determining the size of the protease-resistant fragment of $PrP^{Sc}$ is a simple and reproducible way to classify sporadic CJD patients into six groups with shared clinical and pathologic features. The two common alleles of the PrP gene encode either a methionine (M) or valine (V) at position 129. Thus, sporadic CJD manifests as the MM1, MM2, MV2, VV1, or VV2 molecular subtype. Bosque emphasizes that this study points to unsuspected subtlety in the behavior of prions and illustrates the value of autopsy in suspected CJD.*

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## BAEP monitoring: When is change in wave V significant?

James and Husain examined the sensitivity of intraoperative BAEP changes in predicting hearing loss in patients undergoing posterior fossa surgery. During non-cerebellopontine angle (CPA) tumor surgery, hearing loss usually occurred only with permanent loss of wave V, whereas much smaller changes were associated with hearing loss in CPA tumor surgery.

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*The editorial by Loiselle and Nuwer notes that interpretation criteria for BAEP in the operating room clearly must vary depending on the type of case being monitored. Overreliance upon arbitrary warning criteria can be inaccurate and increase patient risk. On the other hand, watching the BAEP waveform deteriorate until just prior to the disappearance of wave V in non-CPA tumor cases (or any case for that matter) before warning the surgeon does not seem acceptable. The single alarm criterion that is frequently used may need to be replaced with a more complex sliding scale of change.*

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## Evolution of migraine headache

Limited data are available on the natural history of migraine, especially when it progresses to transformed migraine. Bigal et al. studied 402 patients with headaches in the early stages of chronification. Most attacks of transformed migraine fill criteria for migraine. Over time, the disease starts to resemble chronic tension-type headache. The phenotype of transformed migraine thus varies according to the stage of the disease.

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## Susceptibility to compulsive dopaminergic drug use in PD

Evans et al. characterized patients with Parkinson disease (PD) using excessive dopaminergic medication despite harmful drug-induced psychomotor effects vs control patients with PD vs healthy controls. Younger age at disease onset, higher novelty-seeking personality traits, depressive symptoms, and alcohol intake identified patients who compulsively used dopaminergic drugs.

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## Parkinson tremor causes pacemaker misfire

Wills et al. report that rate-adaptive cardiac pacemakers, designed to sense exercise, can instead detect parkinsonian tremor, in this case causing severe tachycardia and behavioral abnormalities.

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## Seeing faces: Expression vs identity in Asperger syndrome

Hefter et al. studied the recognition of both facial identity and expression in social developmental disorders such as Asperger syndrome. Perceiving facial expressions did not correlate with recognizing faces, but with perception of emotion via non-facial cues. This supports both the independence of facial identity and expression perception and a multimodal deficit for emotional processing in these patients.

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## Transmission ratio distortion at SMA locus

Botta et al. examined the segregation of the SMN1 alleles during pregnancy in a large series of spinal muscular atrophy families (314 fetuses from healthy carriers). Statistical analysis demonstrated that the proportion of fetuses predicted with SMA is lower than the 25% expected for a recessive disorder, resulting in a reduced transmission rate of the SMN1-mutated alleles.

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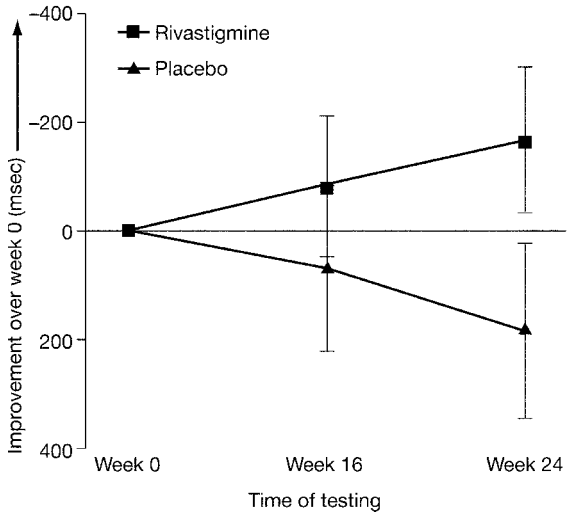
## Rivastigmine, attention, and Parkinson dementia




*Cognitive reaction time.*

Wesnes et al. administered automated attention tests to 487 patients with Parkinson disease (PD) and dementia. Over 24 weeks, rivastigmine, vs placebo and pre-study values, improved each of the four domains of attention that they assessed.

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**November 22 Highlights**

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