

We stress the importance of maintaining a high level of suspicion of PNS (a potentially curable disease) in cases of neurological alterations of unknown cause, once other conditions have been ruled out, even in the absence of onconeural antibodies, since this does not rule out the diagnosis.⁶

Although cases of PNS with no known associated autoantibodies have been described, cases with such complex clinical symptoms are rare. The clinical heterogeneity of our case represents a contribution to the existing knowledge on atypical PNS. Furthermore, it demonstrates the need for studies revealing improvement, which is mandatory in the absence of autoimmune markers.

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Status dissociatus: the most extreme expression of state dissociation[☆]



Status dissociatus: la expresión más extrema de los estados de disociación

Dear Editor,

Status dissociatus (SD) was first described by Mahowald and Schenck¹ in 1991 and is currently considered the most extreme form of state dissociation.² Healthy individuals present 3 states, wakefulness, non-REM (NREM) sleep, and REM sleep,³ each with its own neuroanatomical, neurophysiological, and neurochemical characteristics. In this context, state dissociations result from errors in the normal

process of dynamic changes in the central nervous system as it shifts between states, where one state is interrupted by another (for example, in cataplexy, where the characteristic atonia of the REM sleep appears during wakefulness) or elements of one state persist into the next (for example, sleep paralysis, where wakefulness appears while atonia from the REM sleep state persists).⁴ These errors may occur spontaneously, or as a result of a neurological dysfunction or the use/withdrawal of substances or drugs. These state dissociations should not be confused with dissociative disorders, which are defined in the DSM-V as a group of psychiatric disorders characterised by disruption and/or discontinuation of the normal integration between consciousness, memory, awareness of identity, emotions, perception, body representation, motor control, and behaviour.⁵ The third edition of the International Classification of Sleep Disorders of the American Academy of Sleep Medicine⁶ classifies SD as a subtype of REM sleep behaviour disorder (RBD).

SD is associated with direct thalamic lesions or lesions to structures related to the thalamus, which cause a thalamo-limbic GABAergic dysfunction. It is therefore associated with neurodegenerative diseases (Parkinson's disease or multiple system atrophy), rapidly progressive neurodegenerative diseases (fatal familial insomnia or Creutzfeldt-Jakob disease), autoimmune diseases (autoimmune encephalitis

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or narcolepsy), withdrawal syndromes (delirium tremens), and rare diseases (Morvan or Mulvihill-Smith syndrome).^{6–10} The use of such drugs as antidepressants, cholinesterase inhibitors, or beta-blockers may favour the development of state dissociations in patients with the previously mentioned conditions.

Initial manifestations include a decrease in total sleeping time, hypnagogic hallucinations, or abnormal motor behaviours during sleep, compatible with RSBD. The patient's condition progressively worsens, with the appearance of confusional episodes with hypnagogic hallucinations, eventually progressing to chronic insomnia with motor, verbal, and autonomic hyperactivity, together with lack of circadian rhythm, hallucinations, and dream-like experiences. The most severe stage of SD, with severely disorganised sleep, insomnia, and severe parasomnia, is referred to as *agrypnia excitata*^{11–13} and is considered the final stage, where diseases with direct thalamic involvement converge.

We present the clinical case of a 76-year-old patient who had been referred from the neurology department due to suspicion of sleep apnoea/hypopnoea syndrome; the patient had a medical history of hypertension, glaucoma of the right eye, and akinetic-rigid syndrome, treated with antihypertensive drugs, beta-blockers (eye-drops), and dopamine agonists, respectively. The physical examination revealed gait with short, shuffling steps and no arm swing, and reduced facial expression. A brain MRI scan revealed cortico-subcortical atrophy. In a clinical interview targeting sleep disorder, the patient's wife reported episodes in which he hit and punched her while sleeping, with amnesia after the episode, as well as falling from bed, sleep talking, and confusional arousals. However, the patient and his family's main complaint was excessive daytime sleepiness; after a total of 8–9 h of night-time sleep, he scored 13/21 on the Epworth Sleepiness Scale. A videopolysomnography (v-PSG) study revealed an inability to consolidate sleep, with a severely altered macrostructure; sleep was composed of short periods of NREM sleep (stage N1 sleep exclusively) together with episodes of REM sleep without atonia. We recorded no slow-wave sleep (stage N3) or waves characteristic of stage N2 NREM sleep. These recordings were accompanied by continuous motor hyperactivity, sleep talking, confusion, and disorientation. Motor hyperactivity consisted of simple flexion-extension movements of all limbs, and continuous changes in posture. The electrocardiography channel showed multiple episodes of atrial fibrillation, which had not previously been diagnosed ([online additional material](#)).

The following morning, the patient reported restorative sleep. He was finally diagnosed with intermediate SD, meeting all diagnostic criteria for the condition: very scarce or abnormal sleep stages in the v-PSG (stage N1 and REM), continuous motor hyperactivity, decreased level of consciousness, and lack of circadian rhythm²; the condition was bordering on classically defined SD, characterised by the complete absence of sleep stages.

Studies of akinetic-rigid syndrome have described progression from common state dissociations, such as RSBD

or hypnagogic hallucinations, to SD, which is considered the final stage of the neurodegenerative process. Due to the limited literature on SD, we consider it extremely important to deepen our understanding of its aetiology and progression, as well as its temporal association with concomitant diseases, which would constitute a significant contribution to the monitoring and assessment of these diseases.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at [doi:10.1016/j.jnrleng.2019.02.002](https://doi.org/10.1016/j.jnrleng.2019.02.002).

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New cases of star anise poisoning: are we providing enough information?[☆]

Persisten las intoxicaciones por anís estrellado, ¿estamos dando la suficiente información?

Dear Editor,

Star anise (*Illicium verum* [*I. verum*] Hook. F.) is a plant native to Asia that has been widely used since ancient times for its antioxidant, antimicrobial, expectorant, analgesic, and sedative properties.¹ Since its introduction in Spain, it has been used as carminative, especially for treating baby colic. Despite its long history of use, there continue to be cases of poisoning with neurological and gastrointestinal manifestations, especially in infants younger than 3 months.



We present the case of a 2-month-old patient with no relevant history. He presented episodes of jitteriness in the lower limbs, with increasing frequency over the previous week, displaying startle movements during sleep followed by crying; this had prevented sleeping for more than an hour in the previous 2 days. The patient partially rejected food (baby formula) and presented isolated vomiting with normal bowel movements. He lived in a non-infectious environment and was not taking any medication. Baseline general physical and neurological examinations yielded normal results. We requested a blood analysis including ion concentration, hepatic marker enzymes, and acute-phase reactant levels; results were normal for these parameters and for urine toxicology testing. He was admitted for observation and assessment with other complementary tests.

During admission, we discovered that the mother was giving the patient a star anise tea, which she reported having administered for one week; it was prepared with 1-2 stars in 400-500 cc of boiling water for several minutes.



Figure 1 Resemblance between the fruits of *I. verum* and *I. anisatum*.⁵

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