

isolated extramedullary relapse may be followed by medullar relapse, all authors agree in treating local relapse as a systemic disease, with no consensus on the scheme to be followed. Intrathecal and systemic chemotherapy, and whole brain radiation therapy have been used with regimes that cross the blood-brain barrier, with different results^{2,7}. In summary, when a patient with a history of APL presents neurological symptoms, extramedullary disease should be ruled out, even though the patient is in complete medullar cytological and molecular remission. Prospective studies are needed to determine whether treatment with ATRA *per se* increases the risk of developing extramedullary disease and to assess the need for a study on CSF and CNS chemoprophylaxis upon diagnosis for those at high risk of extramedullary relapse.

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Use of Spanish in scientific publications. Neurology in the context of clinical medicine and Spanish scientific journals

Utilización del español en las publicaciones científicas: la neurología en el contexto de la medicina clínica y las publicaciones científicas españolas

Dear Editor:

It is striking that from some areas such as neurological publications edited in Spain, there has been a defence of the interest and the need to publish in Spanish as opposed to in English¹⁻³, which has become the hegemonic *lingua franca* in the scientific field, especially in the field of natural and biomedical sciences⁴.

To determine the weight and importance of the Spanish language in the scientific production of Spanish institutions in general, and in the area of clinical neurology in particular, as well as the potential impact of publication in Spanish in relation to the degree of citation, a study of the presence of this language in the Science Citation Index-Expanded (SCI-Expanded) has been conducted. The evolution of the impact factor of Spanish biomedical journals has also been studied; this is part of a broader line of research that attempts to characterise the role of non-Anglo-Saxon languages in different areas of knowledge, as well as the

impact of certain publishing strategies related to language, including the translation into English of articles published in other languages.

The analysis of the diachronic evolution of publications (articles, letters, reviews and editorials) containing some SCI-Expanded-included Spanish institutional affiliation between 1979 and 2007 (fig. 1) shows that the language of Cervantes has dropped drastically. This is evidenced by the continued decline found, with values that have not exceeded 10% since the mid-nineties, and that represent only 6.29% of all the works produced by Spanish institutions in 2007. The dramatic reduction in Spanish is a fact, despite the increase in the number of source journals published in Spanish included in this database and, therefore, in the presence of this language in absolute terms. The same declining trend was noted in connection with articles published in journals of the different specialties which make up clinical medicine, even though the presence of Spanish is greater in the area, with values that have remained in any event below 20% since 2005 (fig. 1). However, a contrary trend is observed in clinical neurology, where the Spanish language is in relatively good health. Since the late nineties, the annual scientific production (which hovers around 30-40% of the total number of works produced by Spanish institutions) has remained stable (fig. 1). There are two clinical neurology journals published in this language (*Revista de Neurología* and *NEUROLOGÍA*) that were included in SCI-Expanded in 1997 and 2001, respectively; their editorial policy represents an important impulse in the use of Spanish as a language of scientific communication. Theirs is an edito-



Figure 1 Diachronic evolution of the relative weight of Spanish language in the works produced by Spanish institutions listed in the Science Citation Index Expanded database (percentage of works with respect to total scientific production).

rial policy that, on the other hand, has not been a hindrance in relation to another parameter that is usually the most coveted, the impact factor. This important parameter has experienced a slow but steady increase for these two publications since their inclusion in the *Journal Citation Reports* lists and their evolutions show no significant differences in relation to the impact factors of other Spanish biomedical journals that translate their content into English. In this regard, only one publication stands out far above the rest,

and the others are behind the neurological journals, despite presenting their contents in English (fig. 2).

After satisfying the formal quality and content standards^{4,5}, increasing the degree of citation must, with priority over linguistic issues⁶, pass through the adoption of measures such as ensuring simple, fast and open access⁵, providing clear editor articulation of publication purpose and content and attractive presentation thereof⁷, expanding the number of

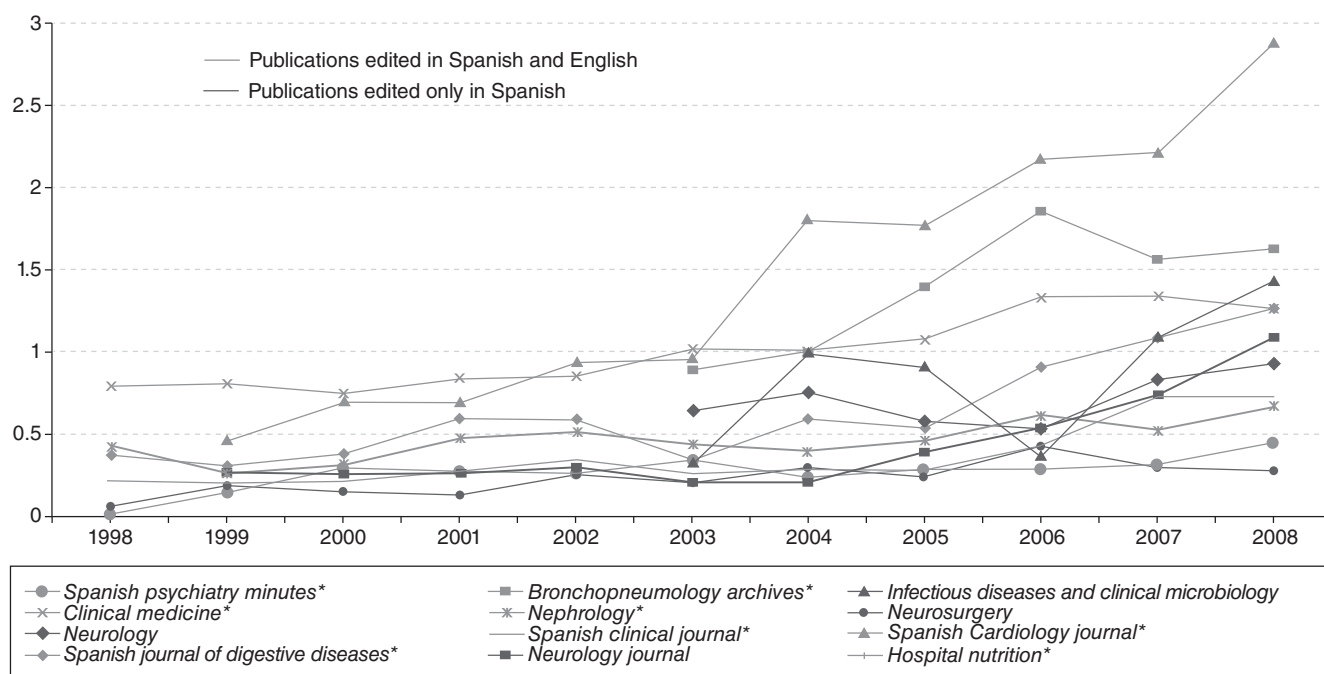


Figure 2 Evolution of the impact factor of biomedical journals published in Spanish included in *Journal Citation Reports*.
*Publications that have an English edition.

authors and encouraging interaction among the networks of authors publishing in the journal⁸, promoting scientific ties with researchers from Spanish-speaking countries⁵, as well as international Spanish researcher collaborations and interdisciplinary connection of clinical research and basic scientific and epidemiologic activity⁹. Furthermore, the policy of prioritising the English language in a publication can lead to unintended and perhaps undesired effects, such as reducing the importance of the publication as a dissemination vehicle for the community which it serves. In this sense, different aspects of great importance have been rightly pointed out from the area. These should be taken into account by all clinically-oriented publications, particularly in a situation characterised by the increasing number of journals indexed in the desired (and at the same time controversial) SSCI-Expanded: namely, that these publications are generally a vehicle of expression for the interests of a scientific society and serve a large community of readers beyond the scientists who publish in them and the role they occupy in the qualifying rankings of publications¹⁰.

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Adult onset of leukoencephalopathy with vanishing white matter

Leucoencefalopatía con sustancia blanca evanescente de inicio en edad adulta

Dear Editor:

We read with great interest the letter published in the previous issue of your journal by Pato Pato et al.¹, which presented a case having characteristics similar to ours. The publication of that case leads us to comment on the possible existence of an underdiagnosis of this disease, which may be more prevalent in the general population and go unnoticed by the entire medical population.

Male, 20 years old, healthy, with no remarkable medical history, son of non-consanguineous parents. In 1994, after a minor traffic accident without head trauma and in which the remaining occupants of the vehicle were unharmed, he presented an episode of psychomotor agitation. Computed tomography (CT) showed bilateral white matter hypodensity, which was initially interpreted as cerebral oedema, requiring mechanical ventilation and admission to the intensive care unit. After extubation nine days later, the patient presented episodes of agitation with echolalia and

palilalia, alternating these with periods during which he was able to maintain simple conversations. He also suffered occasional epileptic crises and right spastic hemiparesis. Brain magnetic resonance imaging (MRI) showed almost complete bilateral affectation of white matter.

The studies performed to rule out common leukoencephalopathy aetiologies were normal: creatine kinase, lactate, protein and immunoglobulins, cerebrospinal fluid analysis, including IgG index, autoimmunity, serology (syphilis, HIV, *Salmonella*, herpes virus), basal cortisol, ACTH, amino acids in urine, very-long-chain fatty acids, analysis of fibroblast culture, urinary arylsulfatase A, phytanic acid, copper and ceruloplasmin, and muscle and sural nerve biopsy.

The patient experienced gradual improvement that became consolidated in the following months; he was able to walk unaided and the epileptic crises disappeared.

Throughout the years of follow-up, he presented progressive deterioration in both motor and cognitive functions, spontaneously or coinciding with fever episodes. Hospitalisation was required in some of them.

The genetic study identified a G338A homozygous mutation in the *eIFGB5* gene. Both parents were carriers of this mutation.

Currently, after 14 years, he is totally dependent in his daily life: he presents marked cognitive impairment, severe