Pulmonary fibrosis: "idiopathic" is not "cryptogenic"

To the Editor:

A recent paper by Wolters *et al.* [1] has raised considerable interest and stirred some debate regarding whether or not the word "idiopathic" should still be part of the name of the disease that we currently call idiopathic pulmonary fibrosis (IPF) [2]. The authors stated that the term idiopathic no longer seemed to describe this progressive lung disease accurately, especially considering the increasing understanding of the causes and pathogenesis of IPF. In a perspective published in the *European Respiratory Journal*, Wells *et al.* [3] responded that IPF remains a truly idiopathic fibrotic disease, with no overall explanation for the development of disease, with the exception of genetic predilection and some familial cases.

I suggest that IPF is indeed idiopathic, although the word idiopathic is generally used somewhat spuriously, and that this debate would gain from an etymological approach. Although in medicine, the word idiopathic is generally used to describe a condition for which no cause has been identified, this usage has somewhat drifted from its etymology. The word idiopathic comes from the ancient Greek ιδιοσ (*idios*, one's own, proper, particular) and πάθος (*páthos*, suffering, *i.e.* disease). Therefore, idiopathic literally means something like "a disease of its own". Although this may often be related to a condition that has no particular cause, the roots are different from those of cryptogenic, from the Greek κρυπτός (hidden) and γένεσις (origin).

IPF has previously been called cryptogenic fibrosing alveolitis in the UK, and it is only recently that the terminology of IPF has been used internationally [4]. Cryptogenic and idiopathic have different roots and slightly different meanings. Based on etymology, the word idiopathic applies better than cryptogenic to what we call IPF, a process with relentless, progressive fibrosis. Although it can be argued that some causes and risk factors of IPF are progressively identified, including tobacco smoking, occupational exposures and genetic risks (and therefore that IPF is not always cryptogenic), pulmonary fibrosis is often compared to abnormal or uncontrolled wound healing, and is indeed a process that progresses "on its own" once initiated [5].

Knowledge of this distinction might help to reconcile the seemingly opposing views on terminology [1, 3] and the use of the word idiopathic in its original sense. In addition, the fact that pulmonary fibrosis is a chronic, progressive and uncontrolled pathogenic process is much better appreciated by patients and relatives than the widely used explanation that idiopathic means that the doctors ignore the cause of this disease, which is a terrible idea for the patient [6].

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"Cryptogenic" and "idiopathic" have different roots and slightly different meanings, which could reconcile the seemingly opposing views on terminology for idiopathic pulmonary fibrosis
http://ow.ly/mvMb30nc2te

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3 Wells AU, Brown KK, Flaherty KR, *et al.* What's in a name? That which we call IPF, by any other name would act the same. *Eur Respir J* 2018; 51: 1800692.
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From the authors:

The writer makes a powerful plea for clarity of thought based on clarity of language, arguing from the Socratic maxim that "the beginning of wisdom is the definition of terms". This etymological perspective is extremely apposite because the distinction between "cryptogenic" and "idiopathic" disease is seminal in the ongoing idiopathic pulmonary fibrosis (IPF) terminology debate.

In our recent perspective [1], we highlighted the dangers of conflating two separate aspects of pathogenesis: the initiation of disease, which may be largely related to epithelial events, and disease progression, which is likely to be influenced by the stromal response that includes fibroblasts and immune cells. Recent advances have largely related to the former and it can be argued, *vide* Cottin, that based on increasing knowledge of predilections and triggers, IPF is less cryptogenic than it once was: perhaps the origin of the disease is now "obscure" but increasingly less "hidden". However, the relentless progression of fibrosis in IPF remains, in the words of Sir Winston Churchill, "a riddle, wrapped in a mystery, inside an enigma": verily, "a disease of its own". We agree that the term "idiopathic" aptly summarises the conundrum that is IPF progression.

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The additional argument advanced by Professor Cottin for retention of the term "idiopathic pulmonary fibrosis" is highly persuasive http://ow.ly/n05630nVCZd

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Reference

1 Wells AU, Brown KK, Flaherty KR, *et al*. What's in a name? That which we call IPF, by any other name would act the same. *Eur Respir J* 2018; 51: 1800692.