



EDITORIAL

Much ado about nothing?

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It all started with the submission of an unsolicited review on idiopathic pulmonary haemosiderosis by IOACHIMESCU *et al.* [1]. During the review process of this excellent manuscript, the idea evolved to launch a whole series on rare (orphan) interstitial lung disorders in the *European Respiratory Journal*. Why bother? Well, admittedly, these diseases are rare, but for the affected patient that does not matter. Patients often feel isolated and like an “orphan” in the field of healthcare themselves [2].

In many of these cases, it takes years from the first symptoms to the definitive diagnosis and initiation of the proper therapy. If cryptogenic organising pneumonia is diagnosed early, corticosteroid therapy may be very beneficial. The same is true for pulmonary Langerhans' cell histiocytosis and smoking cessation, and for alveolar proteinosis and whole-lung lavage. So, from the patient's perspective, it makes a lot of sense to cover these disorders.

In addition, there is the Sherlock Holmes aspect to diagnosis. Currently, there are >200 different entities of diffuse parenchymal lung diseases (DPLDs) [3], some of them familiar to all of us, such as sarcoidosis, hypersensitivity pneumonitis and idiopathic pulmonary fibrosis. Interestingly, even these three most frequent DPLDs would receive the status of an orphan disease according to the definition of a rare disease in Europe, which is “affecting less than one in every 2,000 people” [2]. Besides, there are cases with unusual presentations and findings that may provide confusing leads. Therefore, it can be intellectually challenging to come to a definite diagnosis. Consequently, presentations of rare cases are very popular events at conferences at the national and the international level.

With this as a background, we decided to invite esteemed colleagues to write reviews on rare interstitial lung disorders. The senior authors and the topics covered are as follows:

1) S.R. Johnson: Lymphangioleiomyomatosis; 2) A. Tazi: Adult pulmonary Langerhans' cell histiocytosis; 3) J-F. Cordier: Cryptogenic organising pneumonia; 4) V. Poletti: Diffuse panbronchiolitis; and 5) U. Costabel: Alveolar proteinosis.

The reviews cover historical aspects, pathogenesis, clinical presentation, diagnostic procedures and therapy. The focus of the articles is clinical. The readers should be able to use the manuscript as a guideline for practice. The authors were encouraged to include instructive images, figures and tables. In addition, the reference lists are not limited; they include classical as well as innovative papers.

The first in the series is the article by S.R. Johnson on lymphangioleiomyomatosis in this issue of the *European Respiratory Journal* [4]. One by one, the other articles will follow in subsequent issues. It is hoped that the series will shed some light on this field and help you in your daily work. Nevertheless, tricky cases of rare interstitial lung diseases will always remind us how much truth can be found in the words from *The Importance of Being Earnest* by Oscar Wilde: “The truth is rarely pure and never simple”.

REFERENCES

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