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Review

International consensus: What else can we do to improve diagnosis and therapeutic strategies in patients affected by autoimmune rheumatic diseases (rheumatoid arthritis, spondyloarthritides, systemic sclerosis, systemic lupus erythematosus, antiphospholipid syndrome and Sjogren's syndrome)?
The unmet needs and the clinical grey zone in autoimmune disease management

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ABSTRACT

Autoimmune diseases are a complex set of diseases characterized by immune system activation and, although many progresses have been done in the last 15 years, several unmet needs in the management of these patients may be still identified.

Recently, a panel of international Experts, divided in different working groups according to their clinical and scientific expertise, were asked to identify, debate and formulate a list of key unmet needs within the field of rheumatology, serving as a roadmap for research as well as support for clinicians. After a systematic review of the literature, the results and the discussions from each working group were summarised in different statements. Due to the differences among the diseases and their heterogeneity, a large number of statements was produced and voted by the Experts to reach a consensus in a plenary session. At all the steps of this process, including the initial discussions by the steering committee, the identification of the unmet needs, the expansion of the working group and finally the development of statements, a large agreement was attained.

This work confirmed that several unmet needs may be identified and despite the development of new therapeutic strategies as well as a better understanding of the effects of existing therapies, many open questions still remain in this field, suggesting a research agenda for the future and specific clinical suggestions which may allow physicians to better manage those clinical conditions still lacking of scientific clarity.

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1. Introduction

Autoimmune rheumatologic diseases, pathogenic conditions arising from an abnormal immune response, have been increasingly recognized over the past hundreds of years. The possible causes are not fully understood and both cellular and molecular mechanisms are involved [1,2]. Recently, insights into genetic susceptibility show that environmental triggers may be involved, acting via cellular pathways containing disease-associated polymorphisms. The target tissue provides a decisive microenvironment that affects immune-cell differentiation, leading to a chronic activation of immune system and, thus, development of the autoimmune disease [3,4].

New treatments have been introduced to target different inflammatory pathways and autoimmune rheumatologic diseases. The development of drugs for the treatment of these diseases parallels the increased knowledge of the pathogenic mechanisms. Current treatment guidelines suggest that early diagnosis and initial treatment with immunosuppressive drugs are necessary to limit damage and functional loss and to reduce mortality associated with autoimmune rheumatic disease [5–7]. In this context, it has been shown that frequently the disease course of affected patients is unpredictable as well as their responses, to standard treatments, are variable. Furthermore, it must be pointed out that in many conditions no validated biomarkers exist to predict the course of disease nor the response to therapy.