



Review

The Relapsing Polychondritis Disease Activity Index: Development of a disease activity score for relapsing polychondritis

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ABSTRACT

Objective: The rarity of relapsing polychondritis (RP) has hindered the development of standardized tools for clinical assessment. Here, we describe the development of a preliminary score for assessing disease activity in RP, the Relapsing Polychondritis Disease Activity Index (RPDAI).

Methods: Twenty-seven RP experts participated in an international collaboration. Selection and definition of items for disease activity were established by consensus during a 4-round internet-based Delphi survey. Twenty-six experts assessed the Physician's Global Assessment (PGA) of disease activity on 43 test cases on a 0–100 scale, yielding a total of 1118 PGA ratings. The weight of each item was estimated by

Abbreviations: 95%CI, 95% confidence interval; ARF, acute respiratory failure; GEE, generalized estimating equation; ICC, intraclass correlation coefficient; PGA, Physician's Global Assessment; RP, relapsing polychondritis; RPDAl, Relapsing Polychondritis Disease Activity Index.

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multivariate regression models with generalized estimating equation, using PGA as the dependent variable.

Results: Experts decided in consensus that the RPDAl should consider the 28-day period before each RPDAl assessment. Inter-rater reliability assessed by the intra-class correlation coefficient for the 1118 PGA ratings was 0.51 (CI95%: 0.41–0.64). The final RPDAl score comprised 27 items with individual weights ranging from 1 to 24 and a maximum theoretical RPDAl score of 265. Correlation between the RPDAl scores calculated based on the weights derived from the final multivariate model, and the 1118 PGA ratings was good ($r=0.56$, $p<0.0001$).

Conclusion: We have developed the first consensus scoring system to measure disease activity in relapsing polychondritis (see www.RPDAl.org for online scoring). This tool will be valuable for improving the care of patients with this rare disease.

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1. Introduction

Relapsing polychondritis (RP) is a rare multi-systemic disorder characterized by recurrent, destructive, inflammatory lesions of the auricular, nasal, and laryngo-tracheo-bronchial cartilages [1]. Additional clinical features include ocular inflammation, audio-vestibular impairment, vasculitis, skin involvement, valvular insufficiency, and non-erosive arthritis [2–10]. The rarity of the disease makes it difficult to provide a standardized approach for treatment and follow-up of RP patients, and there is no consensus agreement on any outcome measures in this disease [1]. Standardized disease activity scores would help facilitate the assessment of disease activity in RP, assess the efficacy of novel treatments, and provide prognostic stratification of patients [2]. The lack of such consensual indices has hampered clinical studies in RP and the disease remains an under-researched area. Here, we describe the development and initial validation of a score designed to assess disease activity in RP, the Relapsing Polychondritis Disease Activity Index (RPDAI). This index was developed with the help of a worldwide panel of physicians with significant experience in the care of RP patients. Our main goal was to develop the RPDAl in a manner that this score could be used to standardize disease activity evaluation in RP.

2. Methods

2.1. Expert panel selection

This study reflects a multi-center, international and interdisciplinary collaboration of experts involved in the management of RP, headed by a steering committee composed of an internist specializing in the care of RP (LA) and a fellow in clinical epidemiology (HD). Experts for participation in this study were identified using four sources: (i)

PubMed, searching for lead authors of RP case series published between January 2000 and December 2010; (ii) www.clinicaltrials.gov, searching for principal investigators of current clinical trials in RP; (iii) Board members of European societies of internal medicine and rheumatology, who were contacted for professional referrals; and (iv) French, UK, and US national RP patient associations, who were contacted for personal referrals. This process yielded 37 experts; all were contacted, 29 responded and 27 agreed to participate. Among them were 19 European experts and 8 non-European, their median age was 50 (32–62) years, and the panel included 15 internists, 8 rheumatologists, 2 otolaryngologists, 1 nephrologist and 1 pediatrician. All but 3 (89%) had ≥ 10 years of experience in managing RP patients.

2.2. Preliminary item selection

For the selection and definition of disease activity items, the steering committee prepared a preliminary list grouped by organ system based upon clinical experience [3] and literature review. Eighty-seven items belonging to 10 different domains (constitutional, rheumatologic, chondritis, ophthalmologic, respiratory, otolaryngological, cutaneous, renal, cardiovascular and neurologic) were identified and submitted to the international panel of 27 experts for further selection.

2.3. Delphi survey for item selection

Final item selection was achieved by expert consensus during a four-round internet-based password-protected Delphi survey, a systematic process to derive expert consensus on a topic where the evidence-based data is lacking or scarce [11–13]. Here, all 27 experts rated the importance of each of the 87 preliminary items during four consecutive rounds, and were permitted to suggest new items for

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