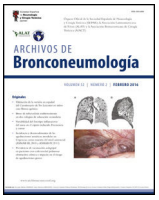
Archivos de Bronconeumología xxx (xxxx) xxx–xxx

Sociedad Española de Neumología y Cirugía Torácica SEPAR

ARCHIVOS DE
Bronconeumología

www.archbronconeumol.org



Editorial

REGINHA—Ibero-American Registry of Hypersensitivity Pneumonitis

The understanding of interstitial lung diseases (ILD) has improved significantly in recent years thanks not only to advances in pathophysiological, diagnostic and therapeutic research, but also to the availability of data from national and international registries on the real-world management of these patients.

Within ILD, hypersensitivity pneumonitis (HP) represents a diagnostic and therapeutic challenge for several reasons: identification of the causal antigen of the disease is difficult; its radiological and histological characteristics are similar to those of other ILDs; and scant evidence has been published on the treatment of this entity.[1–3]

The American Thoracic Society/Japanese Respiratory Society/Latin American Thorax Association (ATS/JRS/ALAT) and CHEST recently issued recommendations for the diagnosis of HP that provide greater diagnostic clarity using the available information. These recommendations are based on the presence of an antigen, tomography findings, bronchoalveolar lavage and histology results, followed by multidisciplinary discussions to attribute different degrees of diagnostic certainty based on these findings.[4–6]

However, despite the diagnostic support provided by the recommendations, diagnostic and therapeutic practices vary greatly among centers worldwide, even among those with expertise in interstitial pathology. Given the low prevalence of ILDs, various registries of patients with these diseases have been implemented to address these issues.

In Latin America, the first continent-wide ILD registry was the Idiopathic Pulmonary Fibrosis Registry (REFIPI). Overall, 146 doctors from 14 countries participated in this study and 761 cases were recorded.[7] The REFIPI reported that the time from the onset of symptoms to diagnosis was 12 months, similar to that published by other registries, and 72% of patients received antifibrotic treatment with pirfenidone or nintedanib. For its part, the Spanish national registry on idiopathic pulmonary fibrosis sponsored by the Spanish Society of Pulmonology and Thoracic Surgery (SEPAR) documented 608 cases from 28 public hospitals. This registry found, among other highly relevant data, that mean time from the onset of symptoms to diagnosis was 20.4 months and that 69.4% of patients received antifibrotic treatment.[8]

Over the years, several national registries have been conducted on ILDs, but the proportion of HP reported in these series is variable and the degree of diagnostic certainty is unclear.[9] In Latin America, an ILD registry conducted in Brazil showed that 740 cases had been exposed to an antigen, but only 327 (44%) were ultimately diagnosed as HP. This disease was the second most common diagnosis (23% of the total) after autoimmune ILD.[10] The results of a Spanish multicenter registry of fibrosing HP that aimed to determine prognostic indicators of progression and mortality have recently been published. After analyzing 103 patients from 12 centers with SEPAR-accredited specialized ILD units, the authors concluded that the presence of fibroblastic foci in a lung biopsy was a predictor of mortality and that the presence of lymphocytosis in BAL was inversely related to mortality. This registry also found that 10% of patients were receiving antifibrotic treatment.[11]

Driven by the experience of REFIPI and the Latin American Registry of Patients with ILD and Myositis-Related Antibodies (EPIMIO), the ILD department of ALAT in collaboration with the ILD division of SEPAR has taken the initiative to set up the Registry of Hypersensitivity Pneumonitis (REGINHA). This is a non-sponsored, bidirectional (prospective and retrospective) registry that includes consecutive cases (alive or deceased) from 2015 onwards in the case of the ALAT protocol, and from 2021 onwards in the case of the SEPAR protocol. The aim is to characterize the clinical, demographic, tomographic, functional, therapeutic and evolutionary characteristics of patients who are diagnosed with HP according to the criteria of the evaluating physician and/or the multidisciplinary group. The registry data reflect the standard clinical practice of each doctor and/or center. The objective of this registry is therefore to determine how HP is diagnosed, how certain the diagnosis is according to the guidelines, and how it is treated in standard clinical practice in Latin America and Spain.

An operational structure has been implemented to conduct this project, and coordinators have been designated in each country to increase dissemination and recruit as many doctors and centers as possible. One representative per center is registered as a user on the RedCap platform after submitting the application form, and is then authorized to enter coded cases into the database in compliance with personal data protection legislation applicable in each country. At least 5 cases with baseline information, a 12-month follow-up, and tomographic images must be uploaded.

REGINHA is already in progress, and we encourage all those interested to join in this project, as this will allow us to better characterize HP at both the diagnostic and therapeutic level in 2 different continents. The data obtained will also help classify patients according to their clinical progress and identify prognostic factors that are involved in this process. The information gathered will be of great use not only to the scientific community but also to our patients.

Please visit the following link to learn more about this project: http://bit.ly/3n3PqMs.

https://doi.org/10.1016/j.arbres.2024.04.025
0300-2896/© 2024 SEPAR. Published by Elsevier España, S.L.U. All rights reserved.

Please cite this article as: M.L. Alberti, E.R. Álvarez and V. Vicens-Zygmunt, REGINHA—Ibero-American Registry of Hypersensitivity Pneumonitis, Archivos de Bronconeumología, https://doi.org/10.1016/j.arbres.2024.04.025

Funding

This study did not receive any type of funding.

Conflict of interests

The authors state that they have no conflict of interests.

References

1. Morell F, Villar A, Montero MÁ, Muñoz X, Colby TV, Pipvath S, et al. Chronic hypersensitivity pneumonitis in patients diagnosed with idiopathic pulmonary fibrosis: a prospective case-cohort study. Lancet Respir Med. 2013;1:685–94.
2. Marinescu DC, Hague CJ, Muller NL, Murphy D, Churg A, Wright JL, et al. Integration and application of radiologic patterns from clinical practice guidelines on idiopathic pulmonary fibrosis and fibrotic hypersensitivity pneumonitis. Chest. 2023;164:1466–75.
3. Alberti ML, Rincon-Alvarez E, Buendia-Roldan I, Selman M. Hypersensitivity pneumonitis: diagnostic and therapeutic challenges. Front Med. 2021;8(September):1–10.
4. Raghu G, Wilson KC, Bargagli E, Bendstrup E, Chami HA, Chua AT, et al. Diagnosis of hypersensitivity pneumonitis in adults: An official ATS/JRS/ALAT clinical practice guideline. Am J Respir Crit Care Med. 2020;202:36–69.
5. Fernández Pérez ER, Travis WD, Lynch DA, Brown KK, Johannson KA, Selman M, et al. Executive summary: diagnosis and evaluation of hypersensitivity pneumonitis: CHEST guideline and expert panel report. Chest. 2021;160:595–615.
6. Buendia-Roldan I, Aguilar-Duran H, Johannson KA, Selman M. Comparing the performance of two recommended criteria for establishing a diagnosis for hypersensitivity pneumonitis. Am J Respir Crit Care Med. 2021;204:865–8.
7. Caro F, Buendía-Roldán I, Noriega-Aguirre L, Alberti ML, Amaral A, Arbo G, et al. Latin American Registry of Idiopathic Pulmonary Fibrosis (REFIPI): clinical characteristics, evolution and treatment. Arch Bronconeumol. 2022;58:794–801.
8. Fernández-Fabrellas E, Molina-Molina M, Soriano JB, Portal JAR, Ancochea J, Valenzuela C, et al. Demographic and clinical profile of idiopathic pulmonary fibrosis patients in Spain: the SEPAR national registry. Respir Res. 2019;20:1–10.
9. Kaul B, Cottin V, Collard HR, Valenzuela C. Variability in global prevalence of interstitial lung disease. Front Med. 2021;8(November):1–10.
10. Lobo S, Matias K, Alberto C, Pereira DC, Raquel M, Castro F, et al. Relative incidence of interstitial lung diseases in Brazil. J Bras Pneumol. 2024;50:1–7.
11. Cano-Jiménez E, Villar Gómez A, Velez Segovia E, Aburto Barrenechea M, Sellarés Torres J, Francesqui J, et al. Prognostic factors of progressive fibrotic hypersensitivity pneumonitis: a large, retrospective, multicenter, observational cohort study. ERJ Open Res. 2024;10:00405–2023.

María Laura Alberti [a,*], Emily Rincón Álvarez [b], Vanesa Vicens-Zygmunt [c]
[a] *Unidad de Enfermedades Pulmonares Intersticiales, Hospital de Rehabilitación Respiratoria "María Ferrer", Ciudad Autónoma de Buenos Aires, Argentina*
[b] *Fundación Neumológica Colombiana, Bogotá, Colombia*
[c] *Unidad de Enfermedades Intersticiales, Hospital Universitari de Bellvitge, Barcelona, Spain*

*Corresponding author.
E-mail address: albertimlaura@gmail.com (M.L. Alberti).