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PROGRESSIVE FIBROSING INTERSTITIAL LUNG DISEASE ASSOCIATED WITH AUTOIMMUNE RHEUMATIC DISEASES: A SNAPSHOT FROM BRAZIL

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BACKGROUND

Interstitial lung disease (ILD) is a common manifestation of autoimmune rheumatic diseases (ARD) and a leading cause of death among these patients¹. A proportion of patients with ARD associated-ILDs (ARD-ILDs) will develop a progressive fibrosing ILD (PF-ILD), characterized by increasing fibrosis on high-resolution computed tomography (HRCT), worsening of lung function, and early mortality². In light of scarce data about the disease in Brazil, we performed a physician survey to explore characteristics of Brazilian PF-ILD patients.

MATERIALS AND METHODS

Forty rheumatologists and forty pulmonologists from Brazil who had managed > 10 non-idiopathic pulmonary fibrosis patients in the past 12 months, and with > 5 and < 35 years of practice were invited to participate in an online interview.

RESULTS

The survey was conducted from December 2019 to January 2020. The participating physicians had an average of 14 years of experience and 7% worked in reference centers. Rheumatologists estimated that ILD developed in 12% of patients with rheumatoid arthritis (RA) and 48% with systemic sclerosis (SSc) compared to 17% of patients with other connective tissue diseases. Interestingly, pulmonologists pointed only 8% of their patients as having SSc. Both specialties estimated that at least 41% of patients with ARD-ILDs develop PF-ILD (Fig. 1). High-resolution computed tomography is the commonest test used to diagnose PF-ILD, mentioned by 100% of physicians, followed by pulmonary function tests (PFTs), used by 85% of rheumatologists and 88% of pulmonologists. On average, patients are seen every 3 months, HRCT is performed every 10 months and PFTs every 7 months. However, pulmonologists perform PFTs every 4.8 months and rheumatologists every 8.3 months, on average. Physicians estimated that, among all ARD-ILDs, patients take 1 year to be diagnosed and those with systemic sclerosis associated-ILD (SSc-ILD) have the fastest evolution, taking 72 months from symptoms to death, while rheumatoid arthritis associated-ILD (RA-ILD) takes 97 months and other connective tissue diseases associated-ILD (other CTD-ILDs) 95 months.

CONCLUSION

Brazilian physicians who manage patients with ARD-ILDs estimate that about half of the patients develop a progressive fibrosing phenotype. Pulmonary function tests and HRCT are the most common tests to diagnose and to follow up these patients. The survey indicated that SSc-ILD has the higher progression rates to death compare with all others ARD-ILD patients.

Figure 1. Estimate percentage of patients with AR-ILDs that develop PF-ILD.

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