SHAREABLE PDF

Pulmonary arterial hypertension associated with primary Sjögren's syndrome: a multicentre cohort study from China

Jieying Wang [1,2,18], Mengtao Li [1,18,19], Qian Wang[1,18], Xiao Zhang[2,18], Junyan Qian[1], Jiuliang Zhao[1], Dong Xu[1], Zhuang Tian[3], Wei Wei[4], Xiaoxia Zuo[5], Miaojia Zhang[6], Ping Zhu[7], Shuang Ye[8], Wei Zhang[9], Yi Zheng[10], Wufang Qi[11], Yang Li[12], Zhuoli Zhang[13], Feng Ding[14], Jieruo Gu[15], Yi Liu[16], Yanhong Wang[17], Yan Zhao[1] and Xiaofeng Zeng[1,19]

Affiliations: [1]Dept of Rheumatology, Peking Union Medical College Hospital, Peking Union Medical College and Chinese Academy of Medical Sciences, National Clinical Research Center for Dermatologic and Immunologic Diseases (NCRC-DID), Key Laboratory of Rheumatology and Clinical Immunology, Ministry of Education, Beijing, China. [2]Dept of Rheumatology, Guangdong Provincial People's Hospital, Guangdong Academy of Medical Sciences, Guangzhou, China. [3]Dept of Cardiology, Peking Union Medical College Hospital, Peking Union Medical College and Chinese Academy of Medical Sciences, Beijing, China. [4]Dept of Rheumatology, Tianjin Medical University General Hospital, Tianjin, China. [5]Dept of Rheumatology, Xiangya Hospital, Central South University, Changsha, China. [6]Dept of Rheumatology, The First Affiliated Hospital of Nanjing Medical University, Nanjing, China. [7]Dept of Clinical Immunology, PLA Specialized Research Institute of Rheumatology and Immunology, Xijing Hospital, Fourth Military Medical University, Xi'an, China. [8]Dept of Rheumatology, Ren Ji Hospital South Campus, School of Medicine, Shanghai Jiao Tong University, Shanghai, China. [9]Dept of Rheumatology, Ren Ji Hospital, School of Medicine, Shanghai Jiao Tong University, Shanghai, China. [10]Dept of Rheumatology, Beijing Chao-Yang Hospital, Capital Medical University, Beijing, China. [11]Dept of Rheumatology, the First Central Hospital, Tianjin, China. [12]Dept of Rheumatology, the Second Affiliated Hospital of Harbin Medical University, Harbin, China. [13]Dept of Rheumatology and Clinical Immunology, Peking University First Hospital, Beijing, China. [14]Dept of Rheumatology, Qilu Hospital of Shandong University, Jinan, China. [15]Department of Rheumatology, the Third Affiliated Hospital of Sun Yat-sen University, Guangzhou, China. [16]Dept of Rheumatology and Immunology, West China Hospital, Sichuan University, Chengdu, China. [17]Dept of Epidemiology and Bio-statistics, Institute of Basic Medical Sciences, China Academy of Medical Sciences & Peking Union Medical College, Beijing, China. [18]J. Wang, M. Li, Q. Wang and X. Zhang contributed equally to this study. [19]M. Li and X. Zeng contributed equally to this article as lead authors and supervised the work.

Correspondence: Xiaofeng Zeng, Dept of Rheumatology, Peking Union Medical College Hospital, No.1 Shuaifuyuan, Beijing 100730, China. E-mail: zengxfpumc@163.com

@ERSpublications
The prognosis of primary Sjögren's syndrome-associated pulmonary arterial hypertension might be improved by improving reserved cardiopulmonary function, by achieving a damage-free state and especially by achieving low-risk category https://bit.ly/3h7mZ9h

Cite this article as: Wang J, Li M, Wang Q, *et al*. Pulmonary arterial hypertension associated with primary Sjögren's syndrome: a multicentre cohort study from China. *Eur Respir J* 2020; 56: 1902157 [https://doi.org/10.1183/13993003.02157-2019].

ABSTRACT
Objectives: Primary Sjögren's syndrome (pSS) is an important cause of pulmonary arterial hypertension (PAH), which remains insufficiently studied and needs attention. This study aimed to investigate the clinical characteristics, risk factors, prognosis and risk assessment of pSS-PAH.

Methods: We established a multicentre cohort of pSS-PAH diagnosed by right heart catheterisation. The case–control study was conducted with pSS-non-PAH patients as a control group to identify the risk factors for PAH. In the cohort study, survival was calculated, and risk assessment was performed at both baseline and follow-up visits.

Results: In total, 103 patients with pSS-PAH were enrolled, with 526 pSS-non-PAH patients as controls. The presence of anti-SSB ($p<0.001$, OR 4.095) and anti-U1RNP antibodies ($p<0.001$, OR 29.518), the age of pSS onset ($p<0.001$, OR 0.651) and the positivity of corneal staining ($p=0.003$, OR 0.409) were identified as independent risk factors for PAH. The 1-, 3- and 5-year survival rates were 94.0%, 88.8% and 79.0%, respectively. Cardiac index ($p=0.010$, hazard ratio (HR) 0.161), pulmonary vascular resistance ($p=0.016$, HR 1.105) and Sjögren's syndrome disease damage index ($p=0.006$, HR 1.570) were identified as potential predictors of death in pSS-PAH. Long-term outcomes were improved in patients in the low-risk category at baseline ($p=0.002$) and follow-up ($p<0.0001$).

Conclusion: The routine screening of PAH is suggested in pSS patients with early onset and positivity for anti-SSB or anti-U1RNP antibodies. Patient prognosis might be improved by improving reserved cardiopulmonary function, by achieving a damage-free state and especially by achieving low-risk category, which supports the treat-to-target strategy for pSS-PAH.