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Clinical Characteristics, Associated Factors, and Mortality-Related Factors of Pneumocystis jirovecii Pneumonia in Patients With Systemic Autoimmune Rheumatic Diseases: A Systematic Review and Meta-analysis

Zhou, LJ; Zhuang, Y; He, X; Zhou, HY; Guo, L; Chen, XX; Ji, JQ.

Corresponding author:
Lijuan Zhou

zlj18200383036@163.com

Author Affiliation:
Department of Rheumatology and Immunology, Sichuan Provincial People's Hospital, School of Medicine, University of Electronic Science and Technology of China, Chengdu, Sichuan, China.

ADMINISTRATIVE INFORMATION

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Review Stage at time of this submission - Data analysis.

Conflicts of interest - None declared.

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Amendments - This protocol was registered with the International Platform of Registered Systematic Review and Meta-Analysis Protocols (INPLASY) on 23 September 2026 and was last updated on 23 September 2026.

INTRODUCTION

Review question / Objective Objective: Pneumocystis jirovecii pneumonia (PJP) is an important opportunistic infection in patients with systemic autoimmune rheumatic diseases (SARDs); however, its clinical characteristics, factors associated with PJP development, and mortality-related factors have not been systematically characterized. This systematic review and meta-analysis aimed to comprehensively evaluate the clinical characteristics of SARD-PJP and identify factors associated with PJP development and mortality.

Condition being studied Systemic autoimmune rheumatic diseases (SARDs) comprise a group of chronic inflammatory disorders characterized by immune system dysregulation. With the widespread use of glucocorticoids, conventional synthetic disease-modifying antirheumatic drugs (csDMARDs), biologic disease-modifying antirheumatic drugs (bDMARDs), and targeted synthetic disease-modifying antirheumatic drugs (tsDMARDs), substantial advances have been made in the management of SARDs. However, while long-term immunosuppressive therapy effectively controls disease activity and delays disease progression, it also increases patients' susceptibility to opportunistic infections. In recent years, accumulating evidence has suggested an increasing incidence of Pneumocystis jirovecii pneumonia (PJP) among patients with SARDs without human immunodeficiency virus (HIV) infection.ARD-PJP often has an insidious onset, progresses rapidly, and is associated with a poor prognosis. The clinical course may be particularly severe in patients with idiopathic inflammatory myopathies (IIM). Therefore, early identification of patients with SARDs at high risk of developing PJP and the establishment of effective risk assessment strategies have become important challenges in clinical management.

Previous studies have suggested that PJP in patients with SARDs is closely associated with high-dose glucocorticoid use, immunosuppressive therapy, and concomitant interstitial lung disease (ILD). However, substantial heterogeneity exists across studies in terms of patient populations, SARD subtypes, and immunosuppressive regimens, and the reported effect sizes of PJP-associated factors have not been entirely consistent.

METHODS

Participant or population Patients with SARDs and PJP.

Intervention None.

Comparator None.

Study designs to be included RCT, prospective, retrospective cohort studies.

Eligibility criteria The inclusion criteria were as follows: (1) the study was a randomized controlled trial, prospective cohort study, or retrospective cohort study; (2) participants fulfilled the relevant classification or diagnostic criteria for systemic autoimmune rheumatic diseases (SARDs), mainly including idiopathic inflammatory myopathies (IIM), systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), systemic sclerosis (SSc), Sjögren's disease (SjD), and ANCA-associated vasculitis (AAV); (3) the study compared the clinical characteristics of patients with SARD-PJP and SARD-non-PJP, and/or evaluated factors associated with PJP development or mortality; and (4) the study provided sufficient data to directly extract or calculate the corresponding effect estimates and their 95% confidence intervals (CIs), including weighted mean differences (WMDs) for continuous variables, odds ratios (ORs) for factors associated with PJP development, and hazard ratios (HRs) for factors associated with mortality.

Information sources PubMed, Embase, Cochrane Library, Web of Science, Scopus, and Ovid databases.

Main outcome(s) Clinical characteristics, factors associated with PJP development, and mortality-related factors.

Quality assessment / Risk of bias analysis Newcastle-Ottawa Scale (NOS).

Strategy of data synthesis Data was analyzed using stata software.All studies that performed pooled analysis were initially tested for heterogeneity using Cochran's Q statistic and inconsistency value (I2). If a p-value of <0.05 or I2≥50% indicated remarkable heterogeneity, a random-effect model and the DerSimonian–Laird (DL) method were ultimately. employed to synthesize the data.

Subgroup analysis Subgroup analyses were performed according to IIM and non-IIM groups.

Sensitivity analysis Sensitivity analyses were performed by the stata software to reflect the sensitivity of an article by the change in the effect size after excluding that article.

Country(ies) involved China.

Keywords systemic autoimmune rheumatic diseases; Pneumocystis jirovecii pneumonia; clinical characteristics; associated factors.

Contributions of each author

Author 1 - Lijuan Zhou.
Email: zlj18200383036@163.com
Author 2 - Yue Zhuang.
Author 3 - Xing He.
Author 4 - Huangyuan Zhou.
Author 5 - Lu Guo.
Author 6 - Xixi Chen.
Author 7 - Jiaqi Ji.