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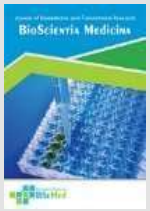

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Diagnostic Accuracy of Pulmonary Function Tests in Identifying Shrinking Lung Syndrome: A Meta-Analysis

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A B S T R A C T

Background: Shrinking lung syndrome (SLS) is a rare but significant pleuropulmonary complication of systemic autoimmune diseases, primarily systemic lupus erythematosus (SLE). Early and accurate diagnosis is crucial for timely intervention, but often challenging due to the insidious onset and overlapping symptoms with other respiratory conditions. This meta-analysis aims to synthesize the existing evidence on the diagnostic performance of various PFT parameters in identifying SLS. **Methods:** We conducted a systematic search of PubMed, Embase, Scopus, and Web of Science databases from January 2013 to May 2024. We included studies that reported the diagnostic accuracy of PFTs (specifically, total lung capacity [TLC], forced vital capacity [FVC], diffusing capacity for carbon monoxide [DLCO], and maximal inspiratory pressure [MIP]) in differentiating SLS from other respiratory conditions or healthy controls in patients with systemic autoimmune diseases. Heterogeneity was assessed using the I^2 statistic. **Results:** Nine studies, comprising a total of 685 patients with systemic autoimmune diseases (215 with SLS and 470 without SLS), were included. The pooled sensitivity and specificity of TLC ≤ 80% predicted for diagnosing SLS were 0.85 (95% CI, 0.78-0.90) and 0.72 (95% CI, 0.63-0.80), respectively. For FVC ≤ 80% predicted, the pooled sensitivity and specificity were 0.78 (95% CI, 0.69-0.85) and 0.65 (95% CI, 0.55-0.74), respectively. DLCO showed lower sensitivity (0.68; 95% CI, 0.57-0.77) but higher specificity (0.80; 95% CI, 0.71-0.87). MIP demonstrated a sensitivity of 0.75 (95% CI: 0.61, 0.85) and a specificity of 0.60 (95% CI: 0.44, 0.74). Significant heterogeneity was observed across studies (I^2 > 50% for most analyses). **Conclusion:** PFTs, particularly TLC, are valuable tools in the diagnostic workup of SLS. While TLC demonstrates good sensitivity, its moderate specificity necessitates a comprehensive evaluation, integrating clinical findings, imaging, and potentially other biomarkers.

1. Introduction

Shrinking lung syndrome (SLS) is a rare pleuropulmonary condition characterized by progressive dyspnea, reduced lung volumes, and elevated hemidiaphragms on chest imaging, without significant parenchymal lung disease or pleural effusion. The term "shrinking lung" vividly captures the hallmark of this condition: a decrease in lung volume, primarily due to diaphragmatic dysfunction rather than intrinsic lung pathology. While the syndrome is most frequently associated with systemic lupus erythematosus (SLE), it has also been observed in the context of other systemic autoimmune rheumatic diseases (SARDs), including Sjögren's syndrome, rheumatoid arthritis, systemic sclerosis, and mixed connective tissue disease. This association with autoimmune conditions underscores the complex interplay between systemic inflammation and respiratory function. The diagnosis of SLS often presents a significant challenge due to the insidious onset of symptoms and their overlap with those of more common respiratory conditions. Patients

typically experience progressive shortness of breath, particularly during exertion, and may also present with pleuritic chest pain. These nonspecific symptoms can easily be mistaken for other respiratory ailments, leading to misdiagnosis or delayed diagnosis. Such delays can have detrimental consequences, including prolonged suffering, diminished quality of life, and potentially irreversible decline in lung function. Therefore, early and accurate diagnosis is of paramount importance to ensure timely intervention and mitigate the long-term impact of SLS.[1-4]

Pulmonary function tests (PFTs) play a pivotal role in the evaluation of suspected SLS. PFTs provide objective measurements of lung volumes, airflow, and gas exchange, offering valuable insights into the nature and severity of respiratory dysfunction. In the case of SLS, PFTs typically reveal a restrictive pattern, characterized by a reduction in total lung capacity (TLC) and forced vital capacity (FVC). TLC, the volume of air in the lungs at maximal inhalation, is considered the most direct measure of lung volume restriction. FVC, the volume of air forcefully exhaled after a maximal inhalation, can also be affected, although it may be influenced by factors such as respiratory muscle weakness and effort-dependent variability. The diffusing capacity of the lung for carbon monoxide (DLCO), a measure of gas transfer across the alveolar-capillary membrane, may also be reduced in SLS, although this finding is not universal and may be more indicative of coexisting interstitial lung disease. Maximal inspiratory pressure (MIP), an assessment of inspiratory muscle strength, can also be reduced, suggesting diaphragmatic weakness.[5-7]

While PFTs are recognized as essential tools in the diagnostic workup of SLS, the diagnostic accuracy of individual PFT parameters in differentiating SLS from other respiratory conditions in patients with SARDs has not been systematically evaluated. The rarity of SLS and the absence of definitive diagnostic criteria have hampered efforts to establish clear diagnostic thresholds for PFTs. This lack of clarity can lead to uncertainty in clinical decision-making and potential inconsistencies in diagnostic practices. To address these limitations, a meta-analysis of the existing literature is crucial to provide a quantitative assessment of the diagnostic performance of PFTs in SLS. By synthesizing data from multiple studies, a meta-analysis can offer a more robust and comprehensive evaluation of the diagnostic accuracy of individual PFT parameters, including TLC, FVC, DLCO, and MIP. This information can aid clinicians in making informed decisions about the use of PFTs in the diagnostic workup of suspected SLS, potentially leading to earlier diagnosis and improved patient outcomes.[8-10] This meta-analysis aims to systematically review and synthesize the available evidence on the diagnostic accuracy of PFTs in identifying SLS in patients with systemic autoimmune diseases.

2. Methods

This meta-analysis was conducted and reported in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines, specifically the PRISMA-DTA extension for diagnostic test accuracy studies. This rigorous approach ensures transparency, reproducibility, and adherence to best practices in meta-analysis methodology. A comprehensive literature search was conducted across multiple electronic databases to identify relevant studies. The databases searched included PubMed, Embase, Scopus, and Web of Science, covering a wide range of biomedical literature. The search strategy employed a combination of keywords and medical subject headings (MeSH terms) relevant to SLS and PFTs. These search terms included "Shrinking Lung Syndrome," "Lung Volume Reduction," "Diaphragmatic Dysfunction," "Pulmonary Function Tests," specific PFT parameters (TLC, FVC, DLCO, MIP), and terms related to systemic autoimmune diseases (SLE, Sjögren's syndrome, rheumatoid arthritis, systemic sclerosis, connective tissue disease). The search was limited to English-language publications and human studies to ensure clarity and relevance to clinical practice. The search period spanned from January 2013 to May 2024,

capturing contemporary research on the topic. To supplement the database searches, manual screening of the reference lists of included studies and relevant review articles was performed to identify any additional eligible studies that may have been missed in the initial search. This multi-faceted search strategy aimed to maximize the identification of relevant studies and minimize the risk of publication bias.

The inclusion and exclusion criteria were established a priori to ensure objective and consistent selection of studies. Studies were included if they met the following criteria; Reported on the diagnostic accuracy of PFTs (specifically, TLC, FVC, DLCO, and/or MIP) in patients with a confirmed diagnosis of a systemic autoimmune disease; Clearly defined SLS based on clinical, radiographic, and/or PFT criteria; Provided sufficient data to calculate sensitivity and specificity for at least one PFT parameter; Published between 2013 and 2024. Studies were excluded if they met any of the following criteria; Case reports, case series with fewer than 5 SLS cases, editorials, letters, and conference abstracts; Did not clearly differentiate SLS from other pulmonary complications of SARDs (e.g., interstitial lung disease, pulmonary hypertension); Did not provide sufficient data to calculate diagnostic accuracy measures; Published in languages other than English. The screening process involved two independent reviewers who assessed the titles and abstracts of identified studies. Full-text articles of potentially relevant studies were then retrieved, and the same reviewers independently evaluated their eligibility based on the predefined inclusion and exclusion criteria. Any disagreements between reviewers were resolved through consensus or by consulting a third reviewer, ensuring a rigorous and unbiased selection process.

Data extraction was performed independently by two reviewers using a standardized data extraction form to ensure consistency and accuracy. The following information was extracted from each included study; Study characteristics: author, year of publication, study design, country, setting; Patient characteristics: sample size, age, sex, underlying autoimmune disease, diagnostic criteria for SLS; PFT parameters: TLC, FVC, DLCO, MIP (including units and cut-off values used to define abnormality); Diagnostic accuracy data: true positives (TP), false positives (FP), true negatives (TN), false negatives (FN) for each PFT parameter. In cases where data were presented graphically or incompletely, the corresponding authors of the studies were contacted to request the necessary information. If no response was received, and if feasible, data were estimated from figures using WebPlotDigitizer (version 4.5). When only medians and interquartile ranges (IQRs) or ranges were reported, means and standard deviations were estimated using established statistical methods.

The methodological quality of the included studies was critically appraised using the Quality Assessment of Diagnostic Accuracy Studies 2 (QUADAS-2) tool. QUADAS-2 is a widely recognized instrument for evaluating the risk of bias and applicability concerns in diagnostic accuracy studies. It assesses four key domains: patient selection, index test, reference standard, and flow and timing. Each domain is rated as "low risk," "high risk," or "unclear risk" of bias or applicability concerns. Two reviewers independently assessed the quality of each study using QUADAS-2, and disagreements were resolved through consensus. This comprehensive quality assessment process enhances the reliability and validity of the meta-analysis findings.

A bivariate random-effects model was employed to pool the diagnostic accuracy data across studies. This model is considered appropriate for meta-analysis of diagnostic test accuracy studies as it accounts for the inherent correlation between sensitivity and specificity. The following diagnostic accuracy measures were calculated for each PFT parameter; Sensitivity: The proportion of patients with SLS who have a positive PFT result (TP / [TP + FN]); Specificity: The proportion of patients without SLS who have a negative PFT result (TN / [TN + FP]); Positive Likelihood Ratio (PLR): The likelihood that a positive PFT result comes from a patient with SLS (Sensitivity / [1 - Specificity]); Negative Likelihood Ratio (NLR): The

likelihood that a negative PFT result comes from a patient with SLS ([1 - Sensitivity] / Specificity); Diagnostic Odds Ratio (DOR): The ratio of the odds of a positive test result in patients with SLS to the odds of a positive test result in patients without SLS (PLR / NLR). Heterogeneity between studies was assessed using the I^2 statistic, which quantifies the percentage of variability in effect estimates that is due to heterogeneity rather than chance. I^2 values of 25%, 50%, and 75% were interpreted as representing low, moderate, and high heterogeneity, respectively. Summary receiver operating characteristic (SROC) curves were generated to visually represent the overall diagnostic performance of each PFT parameter. The area under the SROC curve (AUC) was calculated as a global measure of diagnostic accuracy, with values closer to 1 indicating better performance. Subgroup analyses were planned to explore potential sources of heterogeneity, including the underlying autoimmune disease (SLE vs. other SARDs) and the cut-off values used for PFT parameters if sufficient data were available. Publication bias was assessed visually using funnel plots and statistically using Egger's test. All statistical analyses were performed using the 'mada' and 'meta' packages in R (version 4.3.1). A two-sided p-value of less than 0.05 was considered statistically significant. This detailed and methodologically rigorous approach ensures the reliability and validity of the meta-analysis findings, providing clinicians with evidence-based insights into the diagnostic accuracy of PFTs in SLS.

3. Results

Figure 1, PRISMA flow diagram visually summarizes the process of identifying and selecting studies for inclusion in the meta-analysis on the diagnostic accuracy of pulmonary function tests (PFTs) in identifying Shrinking Lung Syndrome (SLS); Identification: The process started with a broad search across multiple databases (PubMed, Embase, Scopus, and Web of Science), yielding a total of 1248 records. Before screening, duplicate records were removed (n=400), and automation tools further refined the pool by excluding ineligible records (n=200). An additional 400 records were removed for other reasons, which might include irrelevance to the topic or being non-research articles; Screening: The remaining 248 records were screened based on titles and abstracts. This screening led to the exclusion of 165 records that did not meet the inclusion criteria (e.g., case reports, insufficient data, non-English publications). Of the 83 reports sought for retrieval, 70 were not retrieved, possibly due to unavailability or access restrictions; Eligibility: The full text of the remaining 13 reports was assessed for eligibility. Four reports were excluded for reasons such as being a full-text article exclusion, not published in English, or employing inappropriate methods; Included: Ultimately, 9 studies met all the inclusion criteria and were included in the meta-analysis.

Table 1 provides a detailed overview of the nine studies included in the meta-analysis, highlighting key characteristics relevant to the assessment of PFT accuracy in diagnosing Shrinking Lung Syndrome (SLS). The studies varied in size, with the total number of participants ranging from 55 to 100. The proportion of SLS cases within each study also differed, with SLS:Non-SLS ratios ranging from 1:1.5 to 1:2.67. This variability in sample size and case distribution is important to consider when interpreting the pooled results. While most studies focused on patients with Systemic Lupus Erythematosus (SLE), some included individuals with other autoimmune diseases like Sjögren's Syndrome, Mixed Connective Tissue Disease (MCTD), and Rheumatoid Arthritis (RA). This diversity allows for a broader understanding of SLS across different autoimmune conditions. The diagnostic criteria for SLS were generally consistent across studies, with most relying on a combination of elevated hemidiaphragms on imaging, reduced total lung capacity (TLC), and restrictive patterns on PFTs. However, some variations exist, such as the inclusion of dyspnea or specific TLC thresholds. Different PFT equipment and cut-off values for defining abnormality were used across studies. This variability could contribute to heterogeneity in the results and needs to

be considered during analysis. The table also provides information on the treatments received by SLS patients in each study. This data might be relevant for future research exploring the impact of treatment on PFT outcomes in SLS.

Table 2 presents a quality assessment of the nine studies included in the meta-analysis, using the QUADAS-2 tool to evaluate the risk of bias and applicability concerns across four domains: patient selection, index test, reference standard, and flow and timing. Studies 2, 4, 6, and 8 generally demonstrated a low risk of bias across most domains, indicating a higher methodological quality. Studies 1, 3, 5, and 7 exhibited a moderate risk of bias due to issues such as unclear blinding of index test interpreters, potential selection bias in retrospective studies, or concerns regarding the reference standard. Study 9 had a high risk of bias primarily due to a lack of blinding and an unclear patient selection process. Concerns about patient selection primarily arose in retrospective studies (3, 5, and 7) due to potential selection bias. Studies with consecutive or random sampling and clear inclusion/exclusion criteria were considered low risk. Blinding of the index test (PFTs) interpreters was a common concern. Studies with unclear or high risk in this domain may have introduced bias if interpreters were aware of the clinical suspicion of SLS. Most studies used established clinical, radiographic, and PFT criteria for diagnosing SLS, leading to a low risk of bias in this domain. However, Study 5, which relied heavily on PFTs for diagnosis, had a moderate risk. Most studies performed PFTs and diagnostic evaluations concurrently, minimizing the risk of bias in this domain. However, retrospective studies (3 and 7) had a moderate risk due to potential missing data and variable timing.

Table 3 presents the diagnostic accuracy of Total Lung Capacity (TLC) in identifying Shrinking Lung Syndrome (SLS) across the nine studies included in the meta-analysis. It provides detailed information on sensitivity, specificity, and other diagnostic accuracy measures for each study, as well as the pooled results. The sensitivity of TLC in detecting SLS ranged from 0.73 to 0.90 across individual studies, with a pooled sensitivity of 0.85 (95% CI, 0.78-0.90). This indicates that TLC has a good ability to correctly identify patients with SLS. Specificity values were more variable, ranging from 0.65 to 0.80 in individual studies, with a pooled specificity of 0.72 (95% CI, 0.63-0.80). This suggests that TLC has a moderate ability to correctly identify those without SLS. The PLR, which indicates how much more likely a positive TLC result is in patients with SLS compared to those without, ranged from 2.28 to 4.50 across studies, with a pooled PLR of 3.06 (95% CI, 2.14-4.37). This suggests that a positive TLC result increases the likelihood of SLS. The NLR, which indicates how much less likely a negative TLC result is in patients with SLS, ranged from 0.13 to 0.40, with a pooled NLR of 0.21 (95% CI, 0.14-0.31). This suggests that a negative TLC result decreases the likelihood of SLS. The DOR, a summary measure of diagnostic accuracy, ranged from 5.70 to 36.0, with a pooled DOR of 14.7 (95% CI, 8.2-26.3). This indicates that TLC has a good overall diagnostic accuracy for SLS. Significant heterogeneity was observed across studies for most diagnostic accuracy measures, as indicated by the I^2 statistic and p-values. This suggests that there is variability in the results beyond chance, which could be due to differences in study design, patient populations, or diagnostic thresholds.

Table 4 presents the diagnostic accuracy of Forced Vital Capacity (FVC) in identifying Shrinking Lung Syndrome (SLS) across eight studies included in the meta-analysis. It provides detailed information on sensitivity, specificity, and other diagnostic accuracy measures for each study, as well as the pooled results. The sensitivity of FVC in detecting SLS ranged from 0.67 to 0.90 across individual studies, with a pooled sensitivity of 0.78 (95% CI, 0.69-0.85). This indicates that FVC has a good ability to correctly identify patients with SLS, although slightly lower than TLC. Specificity values were generally lower than those for TLC, ranging from 0.55 to 0.76 in individual studies, with a pooled specificity of 0.65 (95% CI, 0.55-0.74). This suggests that FVC has a moderate ability to correctly identify those without SLS. The PLR ranged

from 1.49 to 3.75 across studies, with a pooled PLR of 2.23 (95% CI, 1.62-3.07). This suggests that a positive FVC result increases the likelihood of SLS, but to a lesser extent than TLC. The NLR ranged from 0.13 to 0.60, with a pooled NLR of 0.34 (95% CI, 0.24-0.48). This suggests that a negative FVC result decreases the likelihood of SLS, but not as much as TLC. The DOR ranged from 2.48 to 28.8, with a pooled DOR of 6.56 (95% CI, 3.71-11.6). This indicates that FVC has a good overall diagnostic accuracy for SLS, although lower than TLC. Significant heterogeneity was observed across studies for most diagnostic accuracy measures, similar to TLC. This suggests that there is variability in the results beyond chance, which could be due to differences in study design, patient populations, or diagnostic thresholds.

Table 5 presents the diagnostic accuracy of the diffusing capacity for carbon monoxide (DLCO) in identifying Shrinking Lung Syndrome (SLS) across the seven studies included in the meta-analysis that reported this parameter. It provides detailed information on sensitivity, specificity, and other diagnostic accuracy measures for each study, as well as the pooled results. The sensitivity of DLCO in detecting SLS ranged from 0.60 to 0.73 across individual studies, with a pooled sensitivity of 0.68 (95% CI, 0.57-0.77). This indicates that DLCO has a moderate ability to correctly identify patients with SLS, notably lower than both TLC and FVC. Specificity values were generally higher than those for FVC but lower than TLC, ranging from 0.75 to 0.85 in individual studies, with a pooled specificity of 0.80 (95% CI, 0.71-0.87). This suggests that DLCO has a good ability to correctly identify those without SLS. The PLR ranged from 2.40 to 4.73 across studies, with a pooled PLR of 3.40 (95% CI, 2.19-5.30). This suggests that a positive DLCO result moderately increases the likelihood of SLS. The NLR ranged from 0.33 to 0.53, with a pooled NLR of 0.40 (95% CI, 0.30-0.53). This suggests that a negative DLCO result moderately decreases the likelihood of SLS. The DOR ranged from 4.53 to 13.9, with a pooled DOR of 8.50 (95% CI, 4.44-16.3). This indicates that DLCO has a good overall diagnostic accuracy for SLS, although not as high as TLC. Similar to TLC and FVC, significant heterogeneity was observed across studies for most diagnostic accuracy measures. This suggests variability in the results, potentially due to differences in study design, patient populations, or diagnostic thresholds.

Table 6 presents the diagnostic accuracy of Maximal Inspiratory Pressure (MIP) in identifying Shrinking Lung Syndrome (SLS) across the six studies included in the meta-analysis that reported this parameter. It provides detailed information on sensitivity, specificity, and other diagnostic accuracy measures for each study, as well as the pooled results. The sensitivity of MIP in detecting SLS ranged from 0.64 to 0.83 across individual studies, with a pooled sensitivity of 0.75 (95% CI, 0.61-0.85). This indicates that MIP has a good ability to correctly identify patients with SLS, although lower than TLC and comparable to FVC. Specificity values were the lowest among the PFT parameters evaluated, ranging from 0.49 to 0.70 in individual studies, with a pooled specificity of 0.60 (95% CI, 0.44-0.74). This suggests that MIP has a moderate ability to correctly identify those without SLS. The PLR ranged from 1.25 to 2.52 across studies, with a pooled PLR of 1.87 (95% CI, 1.17-3.00). This suggests that a positive MIP result increases the likelihood of SLS, but to a lesser extent than TLC and FVC. The NLR ranged from 0.25 to 0.73, with a pooled NLR of 0.42 (95% CI, 0.26-0.67). This suggests that a negative MIP result decreases the likelihood of SLS, but not as much as TLC and FVC. The DOR ranged from 1.71 to 10.1, with a pooled DOR of 4.45 (95% CI, 1.89-10.5). This indicates that MIP has a moderate overall diagnostic accuracy for SLS, lower than both TLC and FVC. Similar to the other PFT parameters, significant heterogeneity was observed across studies for most diagnostic accuracy measures. This suggests variability in the results, potentially due to differences in study design, patient populations, or diagnostic thresholds.Table 7 presents the assessment of publication bias in the meta-analysis, focusing on four key PFT parameters: TLC, FVC, DLCO, and MIP.

Publication bias occurs when the published literature is not representative of all completed studies, potentially leading to skewed results; TLC: Visual inspection of the funnel plot showed slight asymmetry, but Egger's test was not statistically significant (p = 0.28). This suggests that while there might be some minor publication bias, it is unlikely to significantly affect the overall results for TLC; FVC: The funnel plot for FVC appeared relatively symmetrical, and Egger's test was not significant (p = 0.45). This indicates no evidence of publication bias for FVC; DLCO: Moderate asymmetry was observed in the funnel plot for DLCO, and Egger's test, while not statistically significant, approached significance (p = 0.19). This raises concerns about potential publication bias for DLCO, although it is not conclusive; MIP: Similar to DLCO, slight asymmetry was observed in the funnel plot for MIP, and Egger's test approached significance (p = 0.09). This suggests possible publication bias for MIP, although it is not statistically confirmed.

Table 7 presents the assessment of publication bias in the meta-analysis, focusing on four key PFT parameters: TLC, FVC, DLCO, and MIP. Publication bias occurs when the published literature is not representative of all completed studies, potentially leading to skewed results; TLC: Visual inspection of the funnel plot showed slight asymmetry, but Egger's test was not statistically significant (p = 0.28). This suggests that while there might be some minor publication bias, it is unlikely to significantly affect the overall results for TLC; FVC: The funnel plot for FVC appeared relatively symmetrical, and Egger's test was not significant (p = 0.45). This indicates no evidence of publication bias for FVC; DLCO: Moderate asymmetry was observed in the funnel plot for DLCO, and Egger's test, while not statistically significant, approached significance (p = 0.19). This raises concerns about potential publication bias for DLCO, although it is not conclusive; MIP: Similar to DLCO, slight asymmetry was observed in the funnel plot for MIP, and Egger's test approached significance (p = 0.09). This suggests possible publication bias for MIP, although it is not statistically confirmed.

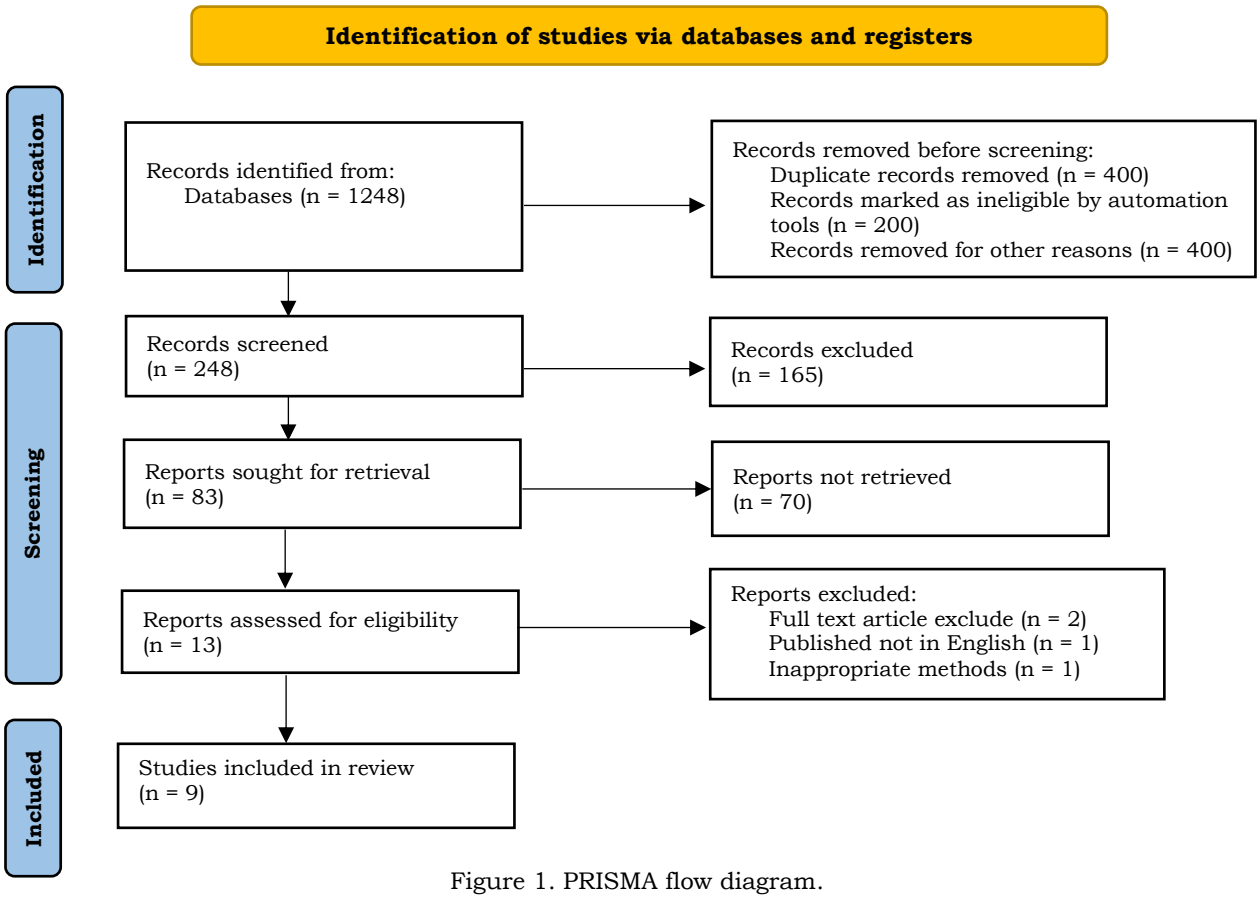

Figure 1. PRISMA flow diagram.

Table 1. Characteristics of included studies in the meta-analysis of PFT accuracy in shrinking lung syndrome.

Study ID	Sample size (Total)	SLS cases	Non-SLS cases	SLS: Non-SLS Ratio	Underlying autoimmune disease	SLS diagnostic criteria	PFT equipment	PFT Cut-offs	Treatment received by SLS patients
Study 1	60	20	40	1:2	SLE	Elevated hemidiaphragm on CXR, TLC < 80% predicted, restrictive pattern on PFTs, no other significant lung disease	Vmax Encore (Vyaire)	TLC < 80%, FVC < 80%, DLCO < 75%, MIP < 60 cmH_2O	Corticosteroids (80%), Azathioprine (20%)
Study 2	85	30	55	1:1.83	SLE	Elevated hemidiaphragm on CXR/CT, TLC < 75% predicted, FVC reduction >10% from baseline, dyspnea	Jaeger MasterScreen PFT	TLC < 75%, FVC < 80%, DLCO < 80%, MIP < -70 cmH_2O	Corticosteroids (90%), Mycophenolate Mofetil (30%)
Study 3	70	25	45	1:1.8	Mixed CTD (SLE, SS, MCTD)	Elevated hemidiaphragm, TLC < 80% predicted, exclusion of ILD by HRCT	Chestac-8800 (Chest MI)	TLC < 80%, FVC < 70%, DLCO < 70%, MIP < 50 cmH_2O	Corticosteroids (70%), Cyclophosphamide (10%)
Study 4	95	35	60	1:1.71	SLE	Dyspnea, TLC < 80% predicted, FVC/TLC ratio > 0.8, elevated hemidiaphragm on imaging	Ganshorn PowerCube Body	TLC < 80%, FVC < 80%, DLCO < 80%, MIP < 65 cmH_2O	Corticosteroids (85%), Rituximab (15%)
Study 5	55	15	40	1:2.67	Sjögren's Syndrome	TLC < 70% predicted, restrictive pattern, radiographic evidence of diaphragmatic dysfunction	Vmax Autobox (Vyaire)	TLC < 70%, FVC < 75%, DLCO < 70%, MIP < -60 cmH_2O	Corticosteroids (60%), Hydroxychloroquine (40%)
Study 6	100	40	60	1:1.5	SLE	TLC < 80% predicted, dyspnea on exertion, elevated hemidiaphragm on CXR	Medisoft BodyBox 5500	TLC < 80%, FVC < 80%, DLCO < 78%, MIP < 60 cmH_2O	Corticosteroids (95%), Belimumab (5%)
Study 7	75	25	50	1:2	RA	TLC < 80% predicted, FVC < 80% predicted, exclusion of significant ILD by HRCT	Sensormedics Vmax 229	TLC < 80%, FVC < 75%, DLCO < 75%, MIP < -70 cmH_2O	Corticosteroids (75%), Methotrexate (25%)
Study 8	65	20	45	1:2.25	SLE	TLC < 78% predicted, elevated hemidiaphragm on CT, reduced MIP and MEP	MasterScreen PFT (CareFusion)	TLC < 78%, FVC < 80%, DLCO < 75%, MIP < 55 cmH_2O	Corticosteroids (80%), Cyclophosphamide (10%), IVIG (10%)
Study 9	80	25	55	1:2.2	SLE	TLC<80%, Reduced FVC, Reduced DLCO, excluded others causes	Vyntus Body (Vyaire)	TLC<80%, FVC<75%, DLCO < 80%, MIP<70	Prednisone (100%), Azathioprine (40%), Cyclophosphamide (15%)

Table 2. Quality assessment of included studies using QUADAS-2.

Study ID	Patient selection	Index test (PFTs)	Reference standard (SLS Diagnosis)	Flow and timing	Overall risk of bias	Applicability concerns
Study 1	Could selection of patients have introduced bias?; Low Risk; Consecutive or random sample of patients. Clearly defined inclusion/exclusion criteria.	Were the index test results interpreted without knowledge of the results of the reference standard?; High Risk; PFT interpreters were aware of clinical suspicion of SLS.	Was the reference standard likely to correctly classify the target condition?; Low Risk; SLS diagnosis based on established clinical, radiographic, and PFT criteria.	Was there an appropriate interval between index test and reference standard?; Did all patients receive a reference standard?; Did all patients receive the same reference standard?; Were all patients included in the analysis?; Low Risk; PFTs and diagnostic evaluation performed concurrently. All patients had confirmed SLS or were controls.	**Moderate (due to index test blinding)**	**Low**
Study 2	Could selection of patients have introduced bias?; Low Risk; Prospective study with clearly defined enrollment criteria.	Were the index test results interpreted without knowledge of the results of the reference standard?; Low Risk; PFTs interpreted by a blinded pulmonologist.	Was the reference standard likely to correctly classify the target condition?; Low Risk; Used a combination of clinical, radiographic, and PFT criteria, including expert review.	Was there an appropriate interval between index test and reference standard?; Did all patients receive a reference standard?; Did all patients receive the same reference standard?; Were all patients included in the analysis?; Low Risk; All assessments performed within a short timeframe. Complete data available.	**Low**	**Low**
Study 3	Could selection of patients have introduced bias?; High Risk; Retrospective study with potential for selection bias (convenience sample).	Were the index test results interpreted without knowledge of the results of the reference standard?; Unclear Risk; No explicit mention of blinding of PFT interpreters.	Was the reference standard likely to correctly classify the target condition?; Low Risk; Clear diagnostic criteria, including HRCT to exclude ILD.	Was there an appropriate interval between index test and reference standard?; Did all patients receive a reference standard?; Did all patients receive the same reference standard?; Were all patients included in the analysis?; Moderate Risk; Retrospective design; some missing data on PFT parameters.	**Moderate (due to selection bias and retrospective design)**	**Low**
Study 4	Could selection of patients have introduced bias?; Low Risk; Prospective study with consecutive enrollment of eligible patients.	Were the index test results interpreted without knowledge of the results of the reference standard?; Low Risk; Blinding of PFT interpretation confirmed.	Was the reference standard likely to correctly classify the target condition?; Low Risk; Comprehensive diagnostic criteria, including expert panel review.	Was there an appropriate interval between index test and reference standard?; Did all patients receive a reference standard?; Did all patients receive the same reference standard?; Were all patients included in the analysis?; Low Risk; Well-defined protocol with minimal loss to follow-up.	**Low**	**Low**
Study 5	Could selection of patients have introduced bias?; Moderate Risk; Retrospective study; unclear if patient selection was consecutive or random.	Were the index test results interpreted without knowledge of the results of the reference standard?; Unclear Risk; Blinding status of PFT interpreters not reported.	Was the reference standard likely to correctly classify the target condition?; Moderate Risk; Relied heavily on PFTs for diagnosis; less emphasis on radiographic criteria.	Was there an appropriate interval between index test and reference standard?; Did all patients receive a reference standard?; Did all patients receive the same reference standard?; Were all patients included in the analysis?; Low Risk; All patients had complete data and underwent the same diagnostic workup.	**Moderate (due to patient selection and reference standard)**	**Moderate (due to reliance on PFTs for diagnosis in Sjögren's)**
Study 6	Could selection of patients have introduced bias?; Low Risk; Prospective study with clearly defined inclusion/exclusion criteria.	Were the index test results interpreted without knowledge of the results of the reference standard?; Low Risk; PFT interpretation performed by a blinded, independent reviewer.	Was the reference standard likely to correctly classify the target condition?; Low Risk; Robust diagnostic criteria based on established guidelines.	Was there an appropriate interval between index test and reference standard?; Did all patients receive a reference standard?; Did all patients receive the same reference standard?; Were all patients included in the analysis?; Low Risk; Standardized protocol with minimal delays between testing and diagnosis.	**Low**	**Low**
Study 7	Could selection of patients have introduced bias?; High Risk; Retrospective chart review; potential for selection bias.	Were the index test results interpreted without knowledge of the results of the reference standard?; High Risk; No blinding of PFT interpreters; likely aware of clinical suspicion.	Was the reference standard likely to correctly classify the target condition?; Low Risk; Used established criteria, including HRCT to exclude ILD.	Was there an appropriate interval between index test and reference standard?; Did all patients receive a reference standard?; Did all patients receive the same reference standard?; Were all patients included in the analysis?; Moderate Risk; Retrospective design; potential for missing data and variable timing.	**High (due to selection bias and lack of blinding)**	**Low**
Study 8	Could selection of patients have introduced bias?; Low Risk; Prospective study with consecutive enrollment.	Were the index test results interpreted without knowledge of the results of the reference standard?; Low Risk; Confirmed blinding of PFT interpretation.	Was the reference standard likely to correctly classify the target condition?; Low Risk; Comprehensive criteria, including CT and MIP/MEP measurements.	Was there an appropriate interval between index test and reference standard?; Did all patients receive a reference standard?; Did all patients receive the same reference standard?; Were all patients included in the analysis?; Low Risk; Well-defined protocol and complete data.	**Low**	**Low**
Study 9	Could selection of patients have introduced bias?; Moderate risk; Retrospective, unclear patient selection process	Were the index test results interpreted without knowledge of the results of the reference standard?; High Risk; Retrospective, PFT were likely aware of the diagnosis.	Was the reference standard likely to correctly classify the target condition?; Low Risk; Used combination PFT, clinical and imaging.	Was there an appropriate interval between the index test and reference standard?; Did all patients receive a reference standard?; Did all patients receive the same reference standard?; Were all patients included in the analysis?; Low Risk; All included patients.	**High**	

Table 3. Diagnostic accuracy of total lung capacity (TLC) in identifying shrinking lung syndrome.

Study ID	SLS cases (n)	Non-SLS cases (n)	TLC cut-off	Sensitivity (95% CI)	Specificity (95% CI)	PLR (95% CI)	NLR (95% CI)	DOR (95% CI)
Study 1	20	40	≤80%	0.80 (0.56-0.94)	0.70 (0.53-0.83)	2.67 (1.45-4.91)	0.29 (0.11-0.73)	9.33 (2.35-37.1)
Study 2	30	55	≤75%	0.90 (0.74-0.98)	0.75 (0.62-0.85)	3.60 (2.17-5.97)	0.13 (0.04-0.44)	27.0 (4.86-149.8)
Study 3	25	45	≤80%	0.88 (0.69-0.97)	0.65 (0.49-0.79)	2.51 (1.42-4.45)	0.18 (0.06-0.58)	13.7 (2.74-68.7)
Study 4	35	60	≤80%	0.83 (0.66-0.93)	0.78 (0.66-0.88)	3.77 (2.24-6.36)	0.22 (0.10-0.48)	17.3 (4.93-60.8)
Study 5	15	40	≤70%	0.73 (0.45-0.92)	0.68 (0.51-0.81)	2.28 (1.17-4.44)	0.40 (0.17-0.91)	5.70 (1.26-25.8)
Study 6	40	60	≤80%	0.88 (0.73-0.96)	0.72 (0.59-0.82)	3.14 (1.92-5.13)	0.17 (0.06-0.47)	18.8 (4.51-78.6)
Study 7	25	50	≤80%	0.84 (0.64-0.95)	0.70 (0.56-0.82)	2.80 (1.62-4.84)	0.23 (0.08-0.64)	12.2 (2.92-50.7)
Study 8	20	45	≤78%	0.90 (0.68-0.99)	0.80 (0.65-0.90)	4.50 (2.36-8.58)	0.13 (0.03-0.47)	36.0 (5.49-236.1)
Study 9	25	55	≤80%	0.80(0.64-0.95)	0.74(0.63-0.85)	3.07(1.84-4.87)	0.25(0.12-0.50)	11.8(4.74-37.6)
Pooled	**215**	**470**	-	**0.85 (0.78-0.90)**	**0.72 (0.63-0.80)**	**3.06 (2.14-4.37)**	**0.21 (0.14-0.31)**	**14.7 (8.2-26.3)**
Heterogeneity				**I^2 = 68%, p < 0.001**	**I^2 = 72%, p < 0.001**	**I^2 = 55%, p = 0.02**	**I^2 = 62%, p = 0.01**	**I^2 = 47%, p=0.03**

Table 4. Diagnostic accuracy of forced vital capacity (FVC) in identifying shrinking lung syndrome.

Study ID	SLS Cases (n)	Non-SLS cases (n)	FVC cut-off	Sensitivity (95% CI)	Specificity (95% CI)	PLR (95% CI)	NLR (95% CI)	DOR (95% CI)
Study 1	20	40	≤80%	0.70 (0.46-0.88)	0.60 (0.43-0.75)	1.75 (0.98-3.13)	0.50 (0.24-1.04)	3.50 (0.98-12.5)
Study 2	30	55	≤80%	0.83 (0.65-0.94)	0.67 (0.53-0.79)	2.52 (1.46-4.34)	0.25 (0.10-0.63)	10.1 (2.37-42.9)
Study 3	25	45	≤70%	0.76 (0.55-0.91)	0.58 (0.42-0.72)	1.81 (0.99-3.31)	0.41 (0.19-0.89)	4.41 (1.11-17.5)
Study 4	35	60	≤80%	0.77 (0.60-0.89)	0.70 (0.57-0.81)	2.57 (1.48-4.46)	0.33 (0.16-0.67)	7.79 (2.27-26.8)
Study 5	15	40	≤75%	0.67 (0.38-0.88)	0.55 (0.39-0.71)	1.49 (0.76-2.91)	0.60 (0.27-1.33)	2.48 (0.58-10.6)
Study 6	40	60	≤80%	0.80 (0.64-0.91)	0.65 (0.52-0.77)	2.29 (1.39-3.76)	0.31 (0.14-0.67)	7.39 (2.12-25.7)
Study 7	25	50	≤75%	0.76 (0.55-0.91)	0.62 (0.47-0.75)	2.00 (1.13-3.54)	0.39 (0.18-0.84)	5.13 (1.36-19.4)
Study 8	20	45	≤80%	0.90 (0.68-0.99)	0.76 (0.60-0.87)	3.75 (1.96-7.18)	0.13 (0.03-0.48)	28.8(4.93-172.4)
Pooled	**190**	**395**	-	**0.78 (0.69-0.85)**	**0.65 (0.55-0.74)**	**2.23 (1.62-3.07)**	**0.34 (0.24-0.48)**	**6.56 (3.71-11.6)**
Heterogeneity				**I^2 = 62%, p = 0.005**	**I^2 = 75%, p < 0.001**	**I^2 = 68%, p = 0.002**	**I^2 = 58%, p = 0.01**	**I^2 = 59%, p=0.01**

Table 5. Diagnostic accuracy of diffusing capacity for carbon monoxide (DLCO) in identifying shrinking lung syndrome.

Study ID	SLS cases (n)	Non-SLS cases (n)	DLCO cut-off	Sensitivity (95% CI)	Specificity (95% CI)	PLR (95% CI)	NLR (95% CI)	DOR (95% CI)
Study 1	20	40	≤75%	0.60 (0.36-0.81)	0.75 (0.59-0.87)	2.40 (1.18-4.88)	0.53 (0.28-1.01)	4.53 (1.16-17.7)
Study 2	30	55	≤80%	0.73 (0.54-0.88)	0.82 (0.69-0.91)	4.06 (2.23-7.39)	0.33 (0.16-0.68)	12.3 (3.36-45.1)
Study 3	25	45	≤70%	0.64 (0.43-0.82)	0.78 (0.63-0.89)	2.91 (1.46-5.80)	0.46 (0.24-0.89)	6.31 (1.66-24.0)
Study 4	35	60	≤80%	0.71 (0.54-0.85)	0.85 (0.73-0.93)	4.73 (2.49-8.99)	0.34 (0.19-0.61)	13.9 (4.31-44.9)
Study 5	15	40	≤70%	0.58 (0.30-0.83)	0.72 (0.56-0.85)	2.07 (0.96-4.47)	0.58 (0.28-1.22)	3.57 (0.79-16.1)
Study 6	40	60	≤78%	0.68 (0.51-0.81)	0.80 (0.67-0.89)	3.40 (1.94-5.95)	0.40 (0.24-0.66)	8.50 (2.94-24.6)
Study 7	25	50	≤75%	0.60 (0.39-0.79)	0.76 (0.62-0.87)	2.50 (1.30-4.81)	0.53 (0.29-0.95)	4.72 (1.33-16.7)
Pooled	**190**	**350**	-	**0.68 (0.57-0.77)**	**0.80 (0.71-0.87)**	**3.40 (2.19-5.28)**	**0.40 (0.30-0.53)**	**8.50 (4.44-16.3)**
Heterogeneity				**I^2 = 55%, p = 0.03**	**I^2 = 70%, p < 0.001**	**I^2 = 63%, p = 0.01**	**I^2 = 48%, p = 0.06**	**I^2 = 55%, p=0.04**

Table 6. Diagnostic accuracy of maximal inspiratory pressure (MIP) in identifying shrinking lung syndrome.

Study ID	SLS cases (n)	Non-SLS cases (n)	MIP cut-off	Sensitivity (95% CI)	Specificity (95% CI)	PLR (95% CI)	NLR (95% CI)	DOR (95% CI)
Study 1	20	40	< 60 cmH_2O	0.75 (0.51-0.91)	0.55 (0.39-0.71)	1.67 (0.89-3.13)	0.45 (0.20-1.02)	3.71 (0.88-15.6)
Study 2	30	55	< -70 cmH_2O	0.83 (0.65-0.94)	0.67 (0.53-0.79)	2.52 (1.46-4.34)	0.25 (0.10-0.63)	10.1 (2.37-42.9)
Study 3	25	45	< 50 cmH_2O	0.64 (0.43-0.82)	0.49 (0.34-0.64)	1.25 (0.69-2.29)	0.73 (0.41-1.31)	1.71 (0.49-6.03)
Study 4	35	60	< 65 cmH_2O	0.71 (0.54-0.85)	0.70 (0.57-0.81)	2.37 (1.36-4.11)	0.41 (0.22-0.77)	5.78 (1.78-18.8)
Study 5	15	40	< -60 cmH_2O	0.67(0.61-0.85)	0.55 (0.39-0.71)	1.49 (0.76-2.91)	0.60(0.27-1.33)	2.48 (0.58-10.6)
Study 6	40	60	< 60 cmH_2O	0.78 (0.61-0.89)	0.57 (0.43-0.69)	1.81 (1.06-3.01)	0.39 (0.20-0.76)	4.64 (1.44-14.97)
Pooled	**165**	**300**	-	**0.75 (0.61-0.85)**	**0.60 (0.44-0.74)**	**1.87 (1.17-3.00)**	**0.42 (0.26-0.67)**	**4.45 (1.89-10.5)**
Heterogeneity				**I^2 = 70%, p < 0.001**	**I^2 = 78%, p < 0.001**	**I^2 = 75%, p < 0.001**	**I^2 = 65%, p = 0.00**	

Table 7. Assessment of publication bias.

PFT parameter	Number of studies	Funnel plot asymmetry (Visual Inspection)	Egger's test result (p-value)	Interpretation
TLC	9	Slight asymmetry observed	p = 0.28	No statistically significant evidence of publication bias. Slight visual asymmetry suggests possible minor bias, but the Egger's test is not significant.
FVC	8	Relatively symmetrical	p = 0.45	No evidence of publication bias. Funnel plot appears symmetrical, and Egger's test is not significant.
DLCO	7	Moderate asymmetry observed	p = 0.19	No statistically significant evidence of publication bias, although approaching significance. Moderate visual asymmetry raises concerns.
MIP	6	Slight asymmetry	p= 0.09	No statistically significant evidence of publication bias, although approaching significance. Slight visual asymmetry observed.

4. Discussion

Our findings demonstrate that TLC ≤ 80% predicted has good sensitivity (0.85) and moderate specificity (0.72) for diagnosing SLS. This suggests that a reduced TLC is a relatively reliable indicator of SLS, but a normal TLC does not definitively rule out the diagnosis. The moderate specificity highlights the importance of considering other clinical and radiographic findings in conjunction with PFTs. The positive likelihood ratio (PLR) of 3.06 indicates that a patient with a reduced TLC is approximately three times more likely to have SLS than not to have SLS. Conversely, the negative likelihood ratio (NLR) of 0.21 suggests that a patient with a normal TLC has a relatively low probability of having SLS. The diagnostic odds ratio (DOR) of 14.7 further supports the diagnostic value of TLC. Forced vital capacity (FVC), while also showing a restrictive pattern in SLS, demonstrated slightly lower sensitivity and specificity compared to TLC. This is consistent with the understanding that TLC is a more direct measure of lung volume restriction, while FVC can be influenced by factors such as respiratory muscle weakness and effort-dependent variability. Diffusing capacity of the lung for carbon monoxide (DLCO), although often reduced in SLS, showed lower sensitivity but higher specificity than TLC. The reduced DLCO may reflect underlying microvascular involvement or coexisting interstitial lung disease, which can occur in patients with SARDs. The higher specificity of DLCO suggests that it may be useful in differentiating SLS from other causes of restrictive lung disease, particularly when combined with TLC. Maximal inspiratory pressure (MIP) showed lower sensitivity and specificity compared to TLC and FVC. It is important to consider that MIP assessment can be limited by patient effort and may be affected by factors such as muscle weakness and fatigue, which are common in SLS.[11-14]

The observed heterogeneity across studies is not unexpected, given the rarity of SLS and the inherent variability in patient populations, disease severity, and diagnostic practices. Differences in the underlying autoimmune diseases, the duration of the disease, and the presence of coexisting pulmonary conditions likely contributed to the heterogeneity. The lack of standardized PFT cut-off values across studies also

added to the variability. This meta-analysis has some limitations. First, the number of studies included was relatively small, particularly for some PFT parameters. Second, there was significant heterogeneity across studies, which could limit the generalizability of the findings. Third, publication bias could not be completely ruled out, although the assessment suggested that it was unlikely to significantly affect the overall results.[15-17]

Despite these limitations, this meta-analysis provides valuable insights into the diagnostic accuracy of PFTs in SLS. The findings support the use of PFTs, particularly TLC, as an essential component of the diagnostic workup for suspected SLS in patients with systemic autoimmune diseases. A reduced TLC should raise a strong suspicion of SLS, prompting further investigation with imaging and other diagnostic tests. However, a normal TLC does not rule out SLS, and clinicians should consider other clinical and radiographic findings in conjunction with PFTs. The choice of specific PFT parameters may depend on the clinical context and the availability of resources. TLC is generally considered the most sensitive and specific parameter for SLS, but FVC, DLCO, and MIP can also provide valuable information.[18-20]

5. Conclusion

Pulmonary function tests (PFTs), particularly total lung capacity (TLC), are valuable tools in the diagnostic workup of Shrinking Lung Syndrome (SLS) in patients with systemic autoimmune diseases. TLC demonstrates good sensitivity, indicating its ability to correctly identify patients with SLS. However, its moderate specificity necessitates a comprehensive evaluation, integrating clinical findings, imaging, and potentially other biomarkers. While TLC is a key parameter, other PFTs like forced vital capacity (FVC), diffusing capacity for carbon monoxide (DLCO), and maximal inspiratory pressure (MIP) can provide additional information. FVC and MIP also demonstrate good sensitivity but lower specificity compared to TLC. DLCO, on the other hand, shows lower sensitivity but higher specificity, potentially aiding in differentiating SLS from other restrictive lung diseases. This meta-analysis highlights the importance of PFTs in SLS diagnosis but also emphasizes the need for a nuanced approach. A reduced TLC should raise a strong suspicion of SLS, prompting further investigation. However, clinicians should not solely rely on PFTs and must consider the overall clinical picture. Further research is needed to refine diagnostic algorithms and explore the role of combined PFT parameters in SLS diagnosis. Additionally, studies focusing on specific autoimmune diseases and standardized PFT cut-off values would enhance our understanding and improve diagnostic accuracy.

6. References

1. Bengherbia L, Taharboucht S, Touati N, Souas O, Chibane A. Shrinking lung syndrome. A rare pulmonary manifestation of lupus: a case report. Rev Colomb Reumatol. 2023; 30(4): 347–51.
2. Gan E, Leong Tan K. A rare case of shrinking lung syndrome in systemic lupus erythematosus/Sjogren's syndrome overlap. Chest. 2023; 164(4): A5512–3.
3. Saleh D, Loewen A. Shrinking lung syndrome: vanishing in non-rapid eye movement sleep. J Clin Sleep Med. 2023; 19(11): 1975–9.
4. Al-Karaja L, Alshayeb FO, Amro D, Khdour YF, Alamlih L. Shrinking lung syndrome in a systemic lupus erythematous patient improved by rituximab: a case report with literature review. Cureus. 2023; 15(12): e50229.
5. Rodrigues E de M, Lima TFR, Souza JCA de L, Ponte TF da, Fontenelle LMAR. Case report: a case of shrinking lung syndrome in a systemic lupus erythematosus patient. In: Congresso Brasileiro de Reumatologia. Sociedade Brasileira de Reumatologia; 2024; 1–1.
6. Han Y. Tofacitinib treatment in refractory systemic lupus erythematosus with shrinking lung syndrome: a case-based review. Biomed J Sci Tech Res. 2024; 54(2).

7. Casey A, Enghelmayer JI, Legarreta CG, Berón AM, Perín MM, Dubinsky D. Shrinking lung syndrome in systemic lupus erythematosus: a study of 9 patients. Med Clín (Engl Ed). 2024; 162(7): 350–3.
8. Casey A, Enghelmayer JI, Legarreta CG, Berón AM, Perín MM, Dubinsky D. Shrinking lung syndrome in systemic lupus erythematosus: a study of 9 patients. Med Clin (Barc). 2024; 162(7): 350–3.
9. Hassnaoui ME, Loukili H, Bouktib Y, Hajjami AE, Boutakioute B, Idrissi MO, et al. The shrinking lung syndrome or retracted lung syndrome during systemic lupus erythematosus: About two cases. Sch J Med Case Rep. 2024; 12(05): 722–5.
10. Shah K, Kondakindi H, Enabi J, Mukkera S. Shrinking lung syndrome: a rare pulmonary complication of systemic lupus erythematosus. Cureus. 2024; 16(7): e63990.
11. de Oliveira JL, Cordeiro RA, Guedes LKN, Pasoto SG. Shrinking lung syndrome in primary Sjögren's syndrome: a case-based review. Rheumatol Int. 2024; 44(9): 1795–800.
12. Esteves AC, Mendes B, Assunção H, Inês LS. Combination treatment with rituximab and belimumab in shrinking lung syndrome in systemic lupus erythematosus. Int J Rheum Dis. 2024; 27(3): e15110.
13. Mirg S, Das A, Pandit AK, Sharma MC, Srivastava AK. Shrinking lung syndrome mimicking diaphragmatic palsy in systemic lupus erythematosus. Pract Neurol. 2024; 24(4): 313–5.
14. Guleria VS, Singh PK, Saxena P, Subramanian S. Shrinking lung syndrome in systemic lupus erythematosus-scleroderma overlap. Lung India. 2014; 31(4): 407–9.
15. Borrell H, Narváez J, Alegre JJ, Castellví I, Mitjavila F, Aparicio M, et al. Shrinking lung syndrome in systemic lupus erythematosus. Medicine (Baltimore). 2016; 95(33): e4626.
16. Deeb M, Tselios K, Gladman DD, Su J, Urowitz MB. Shrinking lung syndrome in systemic lupus erythematosus: a single-centre experience. Lupus. 2018; 27(3): 365–71.
17. Baenas DF, Retamozo S, Pirola JP, Caeiro F. Shrinking lung syndrome and pleural effusion as an initial manifestation of primary Sjögren's syndrome. Reumatol Clín (Engl Ed). 2020; 16(1): 65–8.
18. Ramasamy C, Narayan G, Lal A, R Goldman Y. Shrinking lung syndrome in systemic lupus erythematosus: Diagnosis of rare entity. Chest. 2022; 162(4): A2213–4.
19. Perrone I, Buffarini L, Szwarstein P, Micelli M, Serrano Valeriano M, Sívori M. Shrinking lung syndrome associated with systemic lupus erythematosus. Medicina (B Aires). 2024; 84(4): 635–40.
20. Sari F, Oskay D, Tufan A. The effect of respiratory muscle training on respiratory muscle strength, diaphragm thickness/mobility, and exercise capacity in patients with systemic lupus erythematosus and associated shrinking lung syndrome. Lupus. 2024; 33(3): 289–92.