

**Supplementary File 1:** Details on search strategy.

1. Description of information sources

1.1. MEDLINE (PubMed): search done on the 4th of July, 2018. Last update performed on the 15th of January 2019.

1.2. EMBASE: search done on the 4th of July, 2018. Last update performed on the 15th of January 2019.

1.3. Web of Science search done on the 4th of July, 2018. Last update performed on the 15th of January 2019.

2. Full electronic search strategy

2.1. MEDLINE (PubMed):

("Scleroderma, Systemic"[Mesh] OR "systemic sclerosis" OR scleroderma) AND ("Familial Primary Pulmonary Hypertension"[MeSH Terms] OR "pulmonary arterial hypertension" OR "pulmonary hypertension" OR "primary pulmonary hypertension" OR "secondary pulmonary hypertension" OR "PAH") AND ("videomicroscopy" OR "video microscopy" OR capillaroscopy OR "nailfold videocapillaroscopy" OR "nailfold capillaroscopy" OR angioscopy OR "Microscopic Angioscopy"[Mesh])

2.2. EMBASE:

('systemic sclerosis'/exp OR 'systemic sclerosis' OR 'scleroderma'/exp OR scleroderma) AND ('pulmonary arterial hypertension'/exp OR 'pulmonary arterial hypertension' OR 'pulmonary hypertension'/exp OR 'pulmonary hypertension' OR 'primary pulmonary hypertension'/exp OR 'primary pulmonary hypertension' OR 'secondary pulmonary hypertension'/exp OR 'secondary pulmonary hypertension' OR 'PAH'/exp OR 'PAH') AND ('video microscopy' OR 'video microscopy'/exp OR capillaroscopy OR 'capillaroscopy'/exp OR 'nailfold videocapillaroscopy' OR 'nailfold videocapillaroscopy'/exp OR 'nailfold capillaroscopy' OR 'nailfold capillaroscopy'/exp OR 'angioscopy' OR 'angioscopy'/exp)

2.3. Web of Science:

(TS=("scleroderma, systemic" OR "systemic sclerosis" OR scleroderma)) AND (TS=("Familial Primary Pulmonary Hypertension" OR "pulmonary arterial hypertension" OR "pulmonary hypertension" OR "primary pulmonary hypertension" OR "secondary pulmonary hypertension" OR "PAH")) AND (TS=("videomicroscopy" OR video microscopy" OR "capillaroscopy" OR "nailfold videocapillaroscopy" OR "nailfold capillaroscopy" OR "angioscopy" OR "Microscopic Angioscopy"))

No limitation on publication date was added. Language limitations were added during selection based on title of article. Duplicates were manually removed when articles of all databases were put together to form a list. Titles without abstract or full text were excluded. All study designs with the exception of reviews, editorials, abstracts of scientific congresses and case studies (n < 5) were included.

**Supplementary File 2:** Quality assessment of the selected article.

Question 1 (Q1): Was the research question or objective in this paper clearly stated?

Question 2 (Q2): Was the study population clearly specified and defined?

Question 3 (Q3): Was the participation rate of eligible persons at least 50%?

Question 4 (Q4): Were all the subjects selected or recruited from the same or similar populations (including the same time period)? Were inclusion and exclusion criteria for being in the study pre-specified and applied uniformly to all participants?

Question 5 (Q5): Was a sample size justification, power description, or variance and effect estimates provided?

Question 6 (Q6): For the analyses in this paper, were the exposure(s) of interest (i.e. presence of PAH) measured prior to the outcome(s) being measured?

Question 7 (Q7): Was the timeframe sufficient so that one could reasonably expect to see an association between exposure and outcome if it existed?

Question 8 (Q8): For exposures that can vary in amount or level, did the study examine different levels of the exposure as related to the outcome (e.g., categories of exposure, or exposure measured as continuous variable)?

Question 9 (Q9): Were the exposure measures (independent variables) clearly defined, valid, reliable, and implemented consistently across all study participants?

Question 10 (Q10): Was the exposure(s) assessed more than once over time?

Question 11 (Q11): Were the outcome measures (dependent variables) clearly defined, valid, reliable, and implemented consistently across all study participants?

Question 12 (Q12): Were the outcome assessors blinded to the exposure status of participants?

Question 13 (Q13): Was loss to follow-up after baseline 20% or less?

Question 14 (Q14): Were key potential confounding variables measured and adjusted statistically for their impact on the relationship between exposure(s) and outcome(s)?

Article	Q1	Q2	Q3	Q4	Q5	Q6	Q7	Q8	Q9	Q10	Q11	Q12	Q13	Q14	QR
Sulli, 2012 (30)	Y	Y	NA	Y	N	Y	Y	Y	Y	Y	Y	NR	Y	N	<b>GOOD</b>
Avouac, 2017 (31)	Y	Y	NA	Y	N	Y	Y	Y	Y	Y	Y	Y	Y	Y	<b>GOOD</b>
Hofstee, 2009 (32)	Y	Y	NA	Y	N	Y	Y	Y	Y	NA	Y	Y	NA	N	<b>FAIR</b>
Ricciari, 2013 (33)	Y	Y	NA	Y	N	Y	Y	Y	Y	NA	Y	NR	NA	N	<b>FAIR</b>
Corrado, 2017 (34)	Y	Y	NA	Y	N	Y	Y	Y	Y	NA	Y	Y	NA	N	<b>FAIR</b>

Quality Assessment Tool for Observational Cohort and Cross-Sectional Studies, adapted from the US National institutes of Health (NIH), last updated November 2017, available from: URL: <https://www.nhlbi.nih.gov/health-topics/study-quality-assessment-tools>.

Abbreviations used in table: Q: question; Y: yes; N: no; NA: not applicable; NR: not reported.