SUPPLEMENTARY DATA

Supplementary data 1.

Search strategy in PubMed

#1 ("Lung Diseases, Interstitial"[Mesh:NoExp] OR "Alveolitis, Extrinsic Allergic"[Mesh:NoExp] OR "Bird Fancier's Lung"[Mesh] OR "Farmer's Lung"[Mesh] OR "Silo Filler's Disease"[Mesh] OR "Trichosporonosis"[Mesh] OR "Histiocytosis, Langerhans-Cell"[Mesh:NoExp] OR "Eosinophilic Granuloma"[Mesh] OR "Idiopathic Interstitial Pneumonias"[Mesh:NoExp] OR "Idiopathic Pulmonary Fibrosis"[Mesh] OR "Pulmonary Fibrosis"[Mesh:NoExp] OR Interstitial Lung Disease[tw] OR Interstitial Lung Diseases[tw] OR Interstitial Pneumonia[tw] OR Interstitial Pneumonias[tw] OR Interstitial Pneumonitides[tw] OR Interstitial Pneumonitis[tw] OR Diffuse Parenchymal Lung Disease[tw] OR Diffuse Parenchymal Lung Diseases[tw] OR Hypersensitivity Pneumonitis[tw] OR Hypersensitivity Pneumonitides[tw] OR Extrinsic Allergic Alveolitis[tw] OR Extrinsic Allergic Alveolitides[tw] OR Bird Fancier Lung[tw] OR Bird Fanciers Lung[tw] OR Bird Fancier's Lungs[tw] OR Bird breeders lung[tw] OR Bird breeder's lung[tw] OR Budgerigar Fanciers Lung[tw] OR Budgerigar Fancier's Lung[tw] OR Pigeon Breeders Lung[tw] OR Pigeon Breeder's Lung[tw] OR Farmer Lung[tw] OR Farmers Lung[tw] OR Farmer's Lungs[tw] OR Mushroom Workers Lung[tw] OR Mushroom Worker's Lung[tw] OR Silo Filler Disease[tw] OR Silo Fillers Disease[tw] OR Silo Filler's Disease[tw] OR Silo Fillers' Disease[tw] OR Trichosporonosis[tw] OR Trichosporonoses[tw] OR Schuller-Christian Syndrome[tw] OR Schuller-Christian Disease[tw] OR Schueller-Christian Disease[tw] OR Histiocytosis X[tw] OR Generalized Histiocytosis[tw] OR Generalized Histiocytoses[tw] OR Langerhans Cell Histiocytosis[tw] OR Langerhans Cell Histiocytoses[tw] OR Langerhans-Cell Granulomatosis[tw] OR Letterer-Siwe Disease[tw] OR Non-Lipid Reticuloendotheliosis[tw] OR Nonlipid reticuloendotheliosis[tw] OR Systemic Aleukemic Reticuloendotheliosis[tw] OR Eosinophilic Granuloma[tw] OR Eosinophilic Granulomas[tw] OR eosinophilic granulomatosis[tw] OR Fibrocystic Pulmonary Dysplasia[tw] OR Hamman-Rich Disease[tw] OR Pulmonary fibrosis[tw] OR Pulmonary Fibroses[tw] OR Fibrosing Alveolitis[tw] OR Fibrosing Alveolitides[tw] OR Hamman-Rich Syndrome[tw] OR "Cryptogenic Organizing Pneumonia"[Mesh] OR Organizing pneumonia[tw] OR Organizing pneumonias[tw] OR BOOP[tw] OR organising pneumonia[tw] OR organising pneumonitis[tw] OR organizing pneumonitis[tw] OR NSIP[tw] OR Non-specific interstitial pneumonia[tw] OR Nonspecific interstitial pneumonia[tw] OR Nonspecific interstitial pneumonias[tw] OR Non-specific interstitial pneumonitis[tw] OR Nonspecific interstitial pneumonitis[tw] OR UIP[tw] OR Usual interstitial pneumonia[tw] OR Usual interstitial pneumonias[tw] OR Usual Interstitial Pneumonitis[tw] OR lymphocytic interstitial pneumonia[tw] OR lymphocytic interstitial pneumonitis[tw] OR desquamative interstitial pneumonia[tw] OR desquamative interstitial pneumonitis[tw])

#2("Mortality"[Mesh:NoExp] OR "Survival Rate"[Mesh] OR "Hospital Mortality"[Mesh] OR "Cause of Death"[Mesh] OR "Mortality, Premature"[Mesh] OR "Fatal Outcome"[Mesh] OR "Death"[Mesh] OR mortality[MeSH Subheading] OR "Prognosis"[Mesh] OR "Risk Factors"[Mesh] OR Mortality[tw] OR Mortalities[tw] OR Survival rate[tw] OR Survival rates[tw] OR cause of death[tw] OR causes of death[tw] OR Fatal[tw] OR Fatality[tw] OR Fatalities[tw] OR Death[tw] OR Deaths[tw] OR Died[tw] OR Prognosis[tw] OR Risk factor[tw] OR Risk factors[tw])

#3(("Systemic Vasculitis"[Mesh] OR Systemic Vasculitides OR Vasculitides, Systemic OR Vasculitis, Systemic)) OR ("Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis"[Mesh] OR Anti Neutrophil Cytoplasmic Antibody Associated Vasculitis OR ANCA-Associated Vasculitis OR ANCA Associated Vasculitis OR Vasculitis, ANCA-Associated OR Pauci-Immune Vasculitis OR Pauci Immune Vasculitis OR Pauci-Immune Vasculitides OR Vasculitides, Pauci-Immune OR Vasculitis, Pauci-Immune OR ANCA-Associated Vasculitides OR ANCA Associated Vasculitides OR ANCA-Associated Vasculitide OR Vasculitide, ANCA-Associated OR Vasculitides, ANCA-Associated)) OR ("Churg-Strauss Syndrome"[Mesh] OR Churg Strauss Syndrome OR Syndrome, Churg-Strauss OR Vasculitis, Churg-Strauss OR Churg-Strauss Vasculitis OR Vasculitis, Churg Strauss OR Allergic Granulomatous Angiitis OR Allergic Granulomatous Angiitides OR Angiitides, Allergic Granulomatous OR Granulomatous Angiitides, Allergic OR Granulomatous Angiitis, Allergic OR Allergic Angiitis OR Allergic Angiitides OR Angiitides, Allergic OR Angiitis, Allergic OR Angiitis, Allergic Granulomatous)) OR ("Microscopic Polyangiitis"[Mesh] OR Microscopic Polyangiitides OR Polyangiitides, Microscopic OR Polyangiitis, Microscopic)) OR ("Wegener Granulomatosis"[Mesh] OR Granulomatosis, Wegener OR Wegener's Granulomatosis OR Granulomatosis, Wegener's))
#1 and #2 and #3

Search strategy in Embase

No.	Query	Results
#27.	#16 AND #21 AND #26	368
#26.	#15 OR #24 OR #25	326,702
#25.	'case fatality rate'/exp	2,416
#24.	'mortality rate'/exp	53,328
#23.	#16 AND #21	15,750
#22.	#15 AND #16 AND #21	280
#21.	#17 OR #18 OR #19 OR #20	21,769
#20.	'microscopic polyangiitis'/exp	2,988
#19.	'wegener granulomatosis'/exp	13,421
#18.	'churg strauss syndrome'/exp	4,908
#17.	'anca associated vasculitis'/exp	5,805
#16.	#1 OR #2 OR #4 OR #5 OR #7 OR #8 OR #9 OR #10 OR #11	1,470,218
#15.	#12 OR #13 OR #14	277,948
#14.	'survival rate'/exp	239,125
#13.	'standardized mortality ratio'/exp	2,800
#12.	'morality'/exp	36,328
#11.	'interstitial pneumonia'/exp	15,333
#10.	'silica-induced pulmonary fibrosis'/exp	20
#9.	'experimental pulmonary fibrosis'/exp	1,471
#8.	'fibrosing alveolitis'/exp	23,681
#7.	'lung fibrosis'/exp	78,879
#6.	'interstitial pneumonia'/exp	15,333
#5.	'allergic pneumonitis'/exp	11,419
#4.	'bronchiolitis obliterans organizing	1,829

pneumonia'/exp

#3. 'interstitial pneumonia'/exp 15,333

#2. 'interstitial lung disease'/exp 81,032

#1. 'lung disease'/exp 1,470,218

Search strategy in Cochrane

ID Search Hits

#1 MeSH descriptor: [Antibodies, Antineutrophil Cytoplasmic] explode all trees 69

#2 MeSH descriptor: [Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis] explode all trees 156

#3 MeSH descriptor: [Churg-Strauss Syndrome] explode all trees 27

#4 MeSH descriptor: [Microscopic Polyangiitis] explode all trees 40

#5 MeSH descriptor: [Granulomatosis with Polyangiitis] explode all trees 82

#6 ("Systemic Vasculitis" OR Systemic Vasculitides OR Vasculitides, Systemic OR Vasculitis, Systemic) OR ("Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis" OR Anti Neutrophil Cytoplasmic Antibody Associated Vasculitis OR ANCA-Associated Vasculitis OR ANCA Associated Vasculitis OR Vasculitis, ANCA-Associated OR Pauci-Immune Vasculitis OR Pauci Immune Vasculitis OR Pauci-Immune Vasculitides OR Vasculitides, Pauci-Immune OR Vasculitis, Pauci-Immune OR ANCA-Associated Vasculitides OR ANCA Associated Vasculitides OR ANCA-Associated Vasculitide OR Vasculitide, ANCA-Associated OR Vasculitides, ANCA-Associated) 757

#7 "Churg-Strauss Syndrome" OR Churg Strauss Syndrome OR Syndrome, Churg-Strauss OR Vasculitis, Churg-Strauss OR Churg-Strauss Vasculitis OR Vasculitis, Churg Strauss OR Allergic Granulomatous Angiitis OR Allergic Granulomatous Angiitides OR Angiitides, Allergic Granulomatous OR Granulomatous Angiitides, Allergic OR Granulomatous Angiitis, Allergic OR Allergic Angiitis OR Allergic Angiitides OR Angiitides, Allergic OR Angiitis, Allergic OR Angiitis, Allergic Granulomatous 114

#8 "Microscopic Polyangiitis" OR Microscopic Polyangiitides OR Polyangiitides, Microscopic OR Polyangiitis, Microscopic 185

#9 "Wegener Granulomatosis" OR Granulomatosis, Wegener OR Wegener's Granulomatosis OR Granulomatosis, Wegener's 247

#10 #1 or #2 or #3 or #4 or #5 or #6 or #7 or #8 or #9 949

#11 MeSH descriptor: [Lung Diseases] explode all trees 39288

#12 MeSH descriptor: [Lung Diseases, Interstitial] explode all trees 779

#13 MeSH descriptor: [Idiopathic Interstitial Pneumonias] explode all trees 221

#14 MeSH descriptor: [Pulmonary Fibrosis] explode all trees 493

#15 MeSH descriptor: [Idiopathic Pulmonary Fibrosis] explode all trees 277

#16 "lung diseas*" or "intersitial lung disase*" or "idiopathic interstitial pneumonia*" or "pulmonary fibrosis" or "idiopathic pulmonary fibrosis" or "idiopathic interstitial pneumoni*" or "usual interstitial pneumoni*" 1474

#17 #11 or #12 or #13 or #14 or #15 or #16 40181

#18 MeSH descriptor: [Mortality] explode all trees 12664

#19 MeSH descriptor: [Survival Rate] explode all trees 9570

#20 Mortality or 'Cause of Death' or Child Mortality' or 'Fatal Outcome' or 'Fetal Mortality' or 'Hospital Mortality' or Infant Mortality' or 'Maternal Mortality' or 'Mortality, Premature' or

'Survival Rate' 130205
#21 #18 or #19 or #20 130205
#22 #21 and #17 and #10 34

Supplementary data 2.

Excluded studies

Author/year/title/journal/volume/issue/ISSN/excluded reason

1.Doubkova/Autoantibodies in systemic connective tissue disease and ANCA-associated vasculitis, their relationship to interstitial lung diseases and prognosis/Vnitr Lek/63/2/0042-7732/not enough information

2.Homma/2004/Pulmonary fibrosis in myeloperoxidase antineutrophil cytoplasmic antibody-associated vasculitides /Respirology/9/2/1323-7799/Study on ANCA-positive ILD

3.Takahashi/2005/Development of microscopic polyangiitis in patients with chronic airway disease/Lung/183/4/0341-2040/Study on other patterns of pulmonary involvement in AAV patients

4.Shiraki/2007/Prevalence of myeloperoxidase-anti-neutrophil cytoplasmic antibody (MPO-ANCA) in patients with interstitial pneumonia/Nihon Kokyuki Gakkai Zasshi/45/12/1343-3490 /Study on ANCA-positive ILD

5.Chen/2008/Antineutrophil cytoplasmic autoantibody-associated vasculitis in older patients/Medicine (Baltimore)/87/4/0025-7974 /no morality data

6.Foulon/2008/ANCA-associated lung fibrosis: analysis of 17 patients/Respir Med/102/10/0954-6111/Study on ANCA-positive ILD

7.Hervier/2009/Pulmonary fibrosis associated with ANCA-positive vasculitides. Retrospective study of 12 cases and review of the literature/Ann Rheum Dis/68/3/0003-4967/No comparison group

8.Nakabayashi 2009 Classification of clinical subtypes, patient survival, kidney prognosis, and relapse in patients with MPO-ANCA-associated vasculitis: A single-center experience/Modern Rheumatology/19/4/1439-7595/Study on ANCA-positive ILD

9.Nozu/2009/A comparison of the clinical features of ANCA-positive and ANCA-negative idiopathic pulmonary fibrosis patients/Respiration/77/4/0025-7931/Study on ANCA-positive ILD

10.Feng,/2011/Pathological and high resolution CT findings in Churg-Strauss syndrome/Chin Med Sci J/26/1/1001-9294/no morality data

11.Jin/2011/Clinical features of pulmonary involvement in patients with microscopic polyangiitis/Zhonghua Jie He He Hu Xi Za Zhi/34/5/1001-0939/no morality data

12.Miyazaki/2011/Association of occupational dust exposure with myeloperoxidase-anti-neutrophil cytoplasmic antibody (MPO-ANCA)-positive interstitial pneumonia/American Journal of Respiratory and Critical Care Medicine/183/1/1073-449X/Study on ANCA-positive ILD

13.Nakabayashi/2011/Dual myeloperoxidase-antineutrophil cytoplasmic antibody- and antiglomerular basement membrane antibody-positive cases associated with prior pulmonary fibrosis: a report of four cases/Clin Exp Nephrol/15/2/1342-1751/Study on ANCA-positive ILD

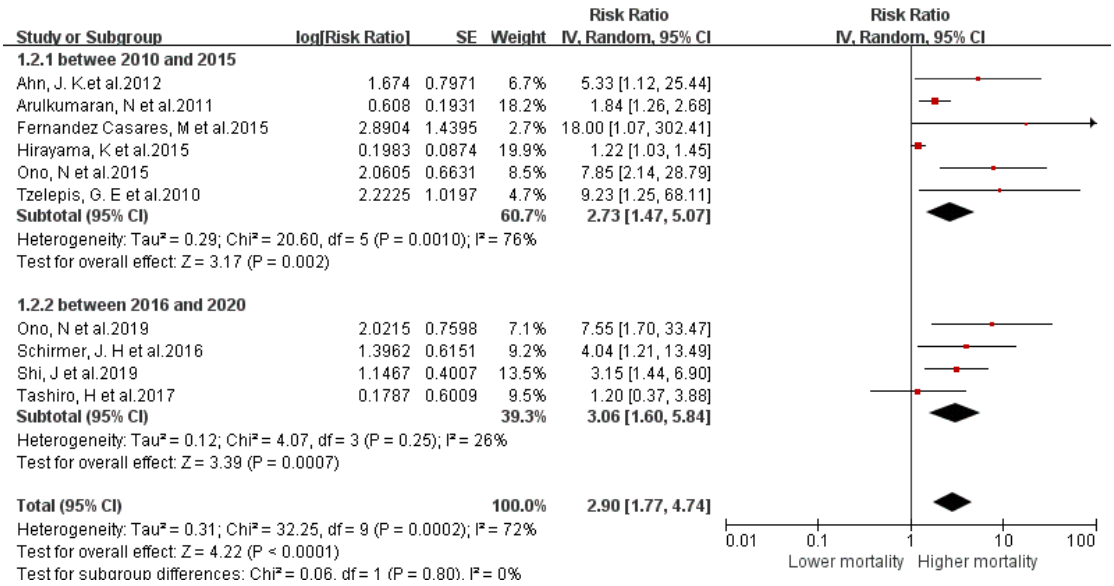
14.Shields/2011/Pyrexia of unknown origin and pulmonary fibrosis as a presentation of MPO-ANCA associated vasculitis/BMJ Case Rep/2011/1757-790x/Not original study

15.Yamada/2011/ANCA: associated lung fibrosis/Semin Respir Crit Care Med/32/3/1069-3424/Not original study
16.Fernandez/2012/Pulmonary fibrosis associated with anti-neutrophil cytoplasmic antibody positive vasculitis/Medicina (B Aires)/72/4/0025-7680/Not original study
17.Ando/2013/Incidence of myeloperoxidase anti-neutrophil cytoplasmic antibody positivity and microscopic polyangitis in the course of idiopathic pulmonary fibrosis/Respir Med/107/4/0954-6111/Study on ANCA-positive ILD
18.Flores/2013/Limited pulmonary MPA, a new MPA entity A rheumatologist's perspective/Clinical and Experimental Nephrology/17/5/1342-1751/Not original study
19.Homma/2013/Pulmonary involvement in ANCA-associated vasculitis from the view of the pulmonologist/Clin Exp Nephrol/17/5/1342-1751/Not original study
20.Sato/2013/Clinical characteristics of interstitial pneumonia in microscopic polyangiitis/Respirology/18 /1323-7799/No comparison group
21.Shoda/2013/Prognosis of MPO-ANCA-positive interstitial pneumonia patients following active treatment/Annals of the Rheumatic Diseases/72/0003-4967/No comparison group
22.Comarmond/2014/Pulmonary fibrosis in ANCA-associated vasculitis: Clinical characteristics and long-term followup of 49 patients/Annals of the Rheumatic Diseases/73/0003-4967/No comparison group
23.Furuta/2014/Comparison of phenotype and outcome in microscopic polyangiitis between Europe and Japan/J Rheumatol/41/2/0315-1623/no morality data
24.Hassan/2014/Lung involvement at presentation predicts disease activity and permanent organ damage at 6, 12 and 24 months follow - up in ANCA - associated vasculitis/BMC Immunology/15/1/1471-2172/no morality data
25.Huang/2014/A retrospective study of microscopic polyangiitis patients presenting with pulmonary fibrosis in China/BMC Pulm Med/14/1471-2466/not enough information
26.Shoda/2014/Prognostic factors for interstitial lung disease with microscopic polyangiitis/Arthritis and Rheumatology/66/2326-5191/not enough information
27.Ishida/2015/Clinical characteristics of combined pulmonary fibrosis and emphysema in patients with connective tissue disease/Annals of the Rheumatic Diseases/74/0003-4967/not enough information
28.Katsumata/2015/Interstitial Lung Disease with ANCA-associated Vasculitis/Clin Med Insights Circ Respir Pulm Med/9 Suppl 1/1179-5484/Not original study
29.Suzuki/2015/Outcomes of and predictive factors in Japanese patients with MPO-ANCA-associated vasculitis: Long-term data from a single rheumatology center/Annals of the Rheumatic Diseases/74/0003-4967/only abstract
30.Yan/2015/Lung performance of anti-neutrophil cytoplasmic antibody (ANCA) associated vasculitis /Respirology/20/1323-7799/only abstract
31.Hozumi/2016/Clinical Implication of Proteinase-3-antineutrophil Cytoplasmic Antibody in Patients with Idiopathic Interstitial Pneumonias/Lung/194/2/0341-2040/Study on ANCA-positive ILD
32.Yamagata/2016/Prevalence and Responsiveness to Treatment of Lung Abnormalities on Chest Computed Tomography in Patients With Microscopic Polyangiitis: A Multicenter, Longitudinal, Retrospective Study of One Hundred Fifty Consecutive Hospital-Based Japanese Patients/Arthritis Rheumatol/68/3/2326-5191/not enough information

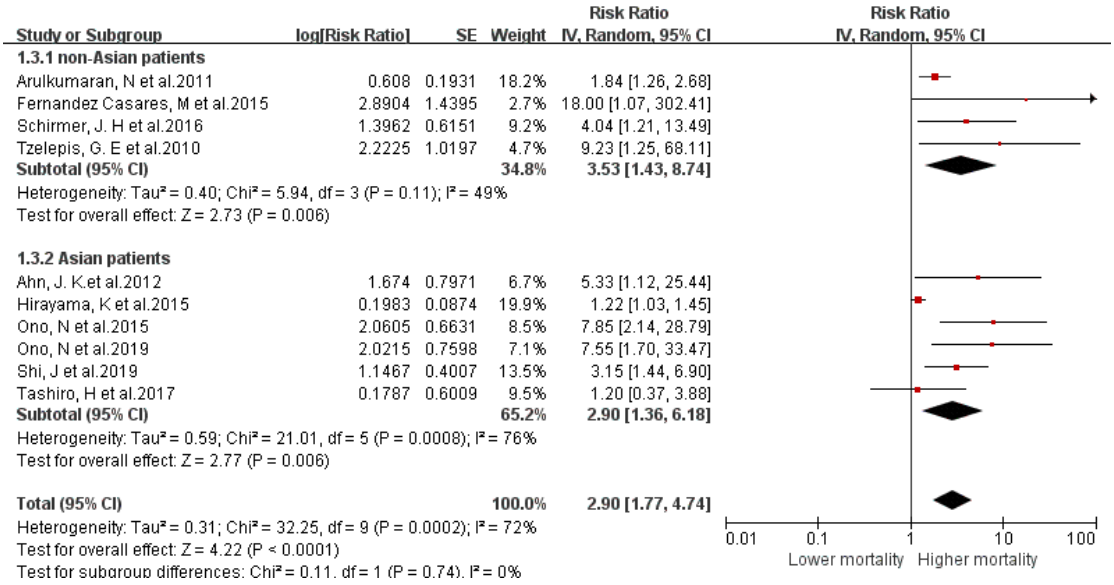
33.Alba/2017/Interstital lung disease in ANCA vasculitis/Autoimmun Rev/16/7/1568-9972/Not original study
34.Mohammad/2017/Pulmonary involvement in antineutrophil cytoplasmic antibodies (ANCA)-associated vasculitis: The influence of ANCA subtype/Journal of Rheumatology/44/10/1499-2752/no morality data
35.Borie/2018/Antineutrophil Cytoplasmic Antibody-Associated Lung Fibrosis/Semin Respir Crit Care Med/39/4/1069-3424/Not original study
36.Garcia/2018/Early interstitial lung disease in microscopic polyangiitis: Case report and literature review/Reumatol Clin/14/2/1699-2584/Not original study
37.Roszkiewicz/2018/From fibrosis to diagnosis: a paediatric case of microscopic polyangiitis and review of the literature/Rheumatol Int/38/4/0172-8172/Not original study
38.Russell/2018/Prognostic Significance of Cavitary Lung Nodules in Granulomatosis With Polyangiitis (Wegener's): A Clinical Imaging Study of 225 Patients/Arthritis Care and Research/70/7/2151-4658/Study on ANCA-positive ILD
39.Shoda/2018/Prognostic factors for interstitial lung disease with microscopic polyangiitis/Annals of the Rheumatic Diseases/77/1468-2060/only abstract
40.Wick/2018 /Pulmonary disorders that are potentially associated with anti- neutrophilic cytoplasmic antibodies: A brief review/Semin Diagn Pathol/35/5/0740-2570/Not original study
41.Baqir/2019/Radiologic and pathologic characteristics of myeloperoxidase-antineutrophil cytoplasmic antibody–associated interstitial lung disease: A retrospective analysis/Sarcoidosis Vasculitis and Diffuse Lung Diseases/36/3/2532-179/no morality data
42.Manfredi/2019/Acute exacerbation of interstitial lung diseases secondary to systemic rheumatic diseases: a prospective study and review of the literature/J Thorac Dis/11/4/2072-1439/Studies comparing ILD-MPA and other ILD patterns
43.Zhao/2019/Clinical features and prognosis of microscopic polyangiitis with usual interstitial pneumonia compared with idiopathic pulmonary fibrosis/Clin Respir J/13/7/1752-6981/No comparison group
44.Maillet/2020/Usual interstitial pneumonia in ANCA-associated vasculitis: A poor prognostic factor/J Autoimmun/106/0896-841/No comparison group.

Supplementary figures

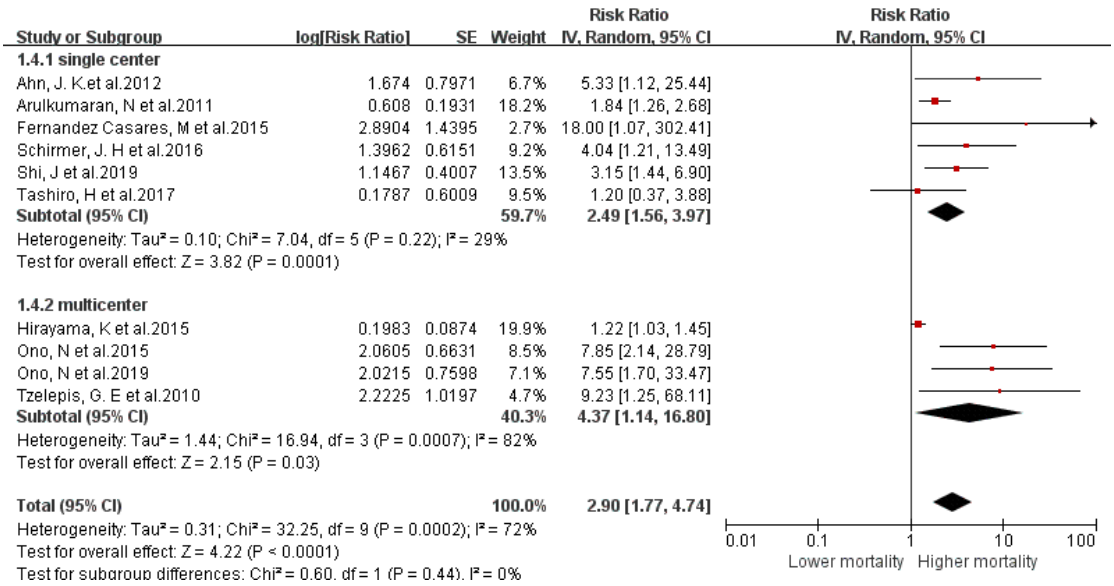
Supplementary figure 1: Subgroup analysis of risk ratio for all-cause mortality according to enrollment period. Enrollment period was classified as 2010 to 2015 and 2016 to 2020.



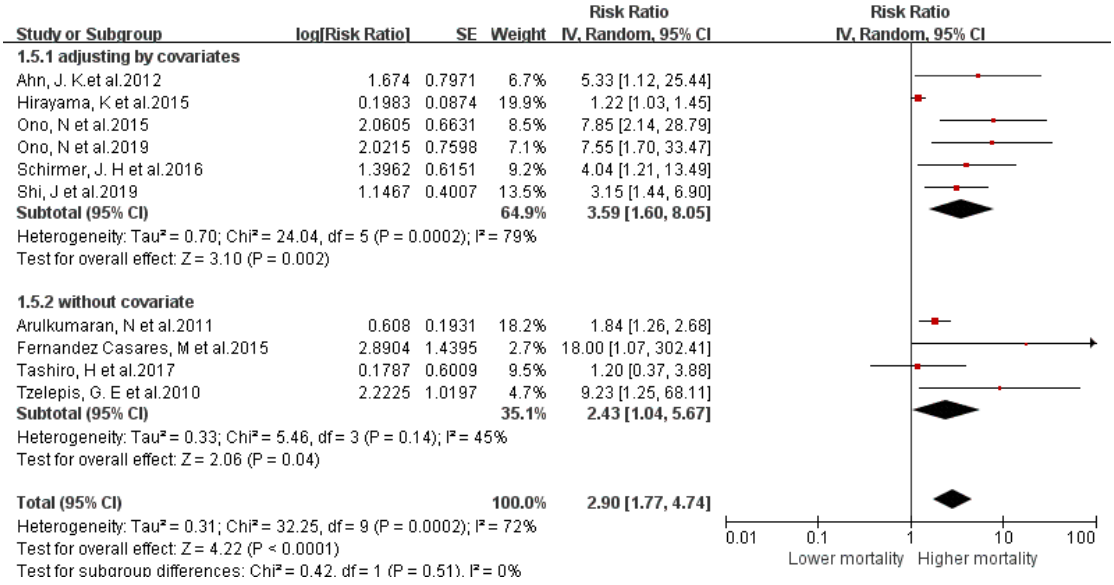
Supplementary figure 2: Subgroup analysis of risk ratio for all-cause mortality according to ethnicity. Ethnicity was classified as Asian patients and non-Asian patients.



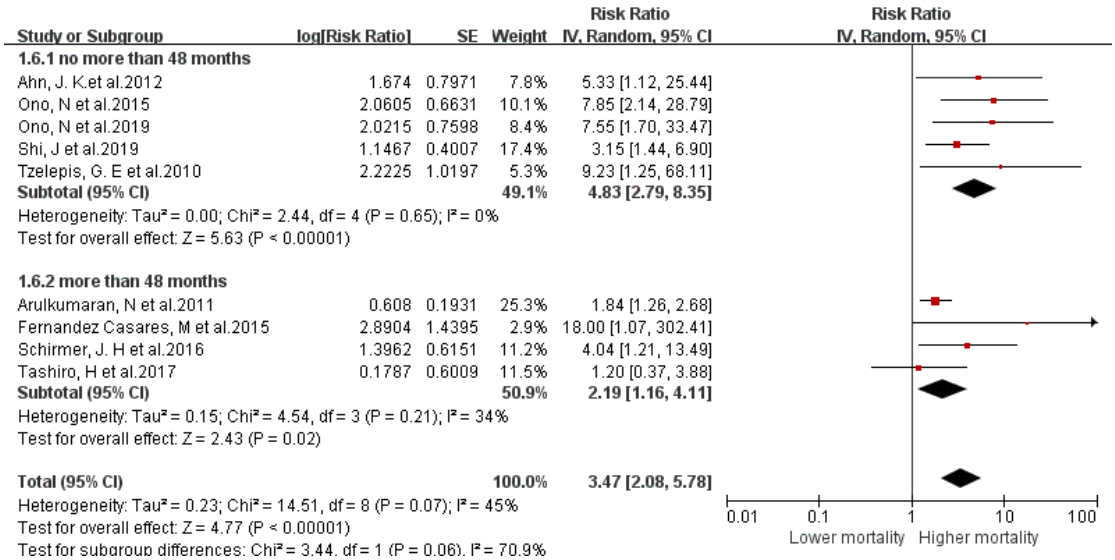
Supplementary figure 3: Subgroup analysis of risk ratio for all-cause mortality according to research center. Research center was classified as single center and multicenter.



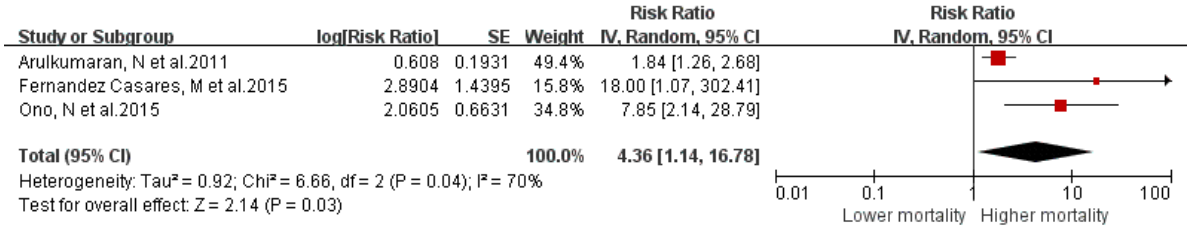
Supplementary figure 4: Subgroup analysis of risk ratio for all-cause mortality according to analysis method. Analysis method was classified as adjusting by covariates and without covariate.



Supplementary figure 5: Subgroup analysis of risk ratio for all-cause mortality according to follow-up period. Follow-up period was classified as no more than 48 months and more than 48 months.



Supplementary figure 6: Subgroup analysis of risk ratio for all-cause mortality according to the pattern of interstitial lung disease. Usual interstitial pneumonia was classified as a specific pattern.



Supplementary figure 7: Subgroup analysis of risk ratio for all-cause mortality according to the use of immunosuppressants for induction treatment.