**e-Table 1 28 potential prognostic factors identified for all-cause mortality**

| |
|---|
| Demographic characteristics |
| age, sex, CADM, dermatomyositis, types of ILD |
| Symptoms |
| fever, cough, dyspnea, muscle weakness |
| Pulmonary function tests |
| %FVC, %DLCO |
| Radiological features |
| GGO/GGA, consolidation, reticular opacity, extent of abnormality |
| Laboratory findings |
| anti-Jo-1 antibody, anti-MDA5 antibody, anti-MDA5 antibody titer, ANA, CK, aldolase, KL-6, LDH, ferritin, A-aDO2, PaO2, CRP, AST |

A-aDO2, alveolar-arterial oxygen difference; ANA, anti-nuclear antibody; anti-MDA5 antibody, anti-melanoma differentiation antigen 5 antibody; AST, asparate aminotransferase; CADM, clinically amyopathic dermatomyositis; CK, creatine kinase; CRP, C-reactive protein; %DLCO, percentage of predicted diffusion capacity of the lung for carbon monoxide; %FVC, percentage of predicted forced vital capacity; GGA, ground glass attenuation; GGO, ground glass opacity; ILD, interstitial lung disease; KL-6, Krebs von den Lungen-6; LDH, lactate dehydrogenase; PaO2, arterial oxygen pressure;

## e-Table 2 Characteristics of 32 studies included in the analysis of potential prognostic factors for all-cause mortality

| Study | Country[a] | Patients (n) | Study design | Follow-up lengths | Subsets of the disease | Types of ILD | Myositis specific antibody (n)[b] |
|---|---|---|---|---|---|---|---|
| Hozumi2018[19] | Japan | 69 | Retrospective cohort | 5.2 (0.2-18.8) years (median (range)) | PM, DM, CADM | Acute/subacute, Chronic | Anti-MDA5 antibody (17)<br>Anti-ARS antibody (29) |
| Kurasawa2018[28] | Japan | 15 | Retrospective cohort | Not specified | DM, ADM | Rapidly progressive | Anti-MDA5 antibody (15) |
| Sugiyama2018[29] | Japan | 116 | Retrospective cohort | Not specified | PM, DM, CADM | Not specified | Anti-MDA5 antibody (8)<br>Anti-ARS antibody (30)[c] |
| Zhang2018[30] | China | 103 | Retrospective cohort | 21.9 (0.5-120) months (mean (range)) | DM | Acute/subacute, Chronic | Anti-Jo-1 antibody (7) |
| Enomoto2017[20] | Japan | 48 | Retrospective cohort | 85.2 (5.8-211) months (median (range)) | PM, DM, CADM | Not specified | Anti-MDA5 antibody (14)<br>Anti-ARS antibody (19) |
| Okabayashi2017[31] | Japan | 14 | Retrospective cohort | Not specified | CADM | Rapidly progressive | Anti-MDA5 antibody (10)<br>Anti-ARS antibody (3)[d] |
| Li2016[25] | China | 26 | Prospective cohort | Not specified | CADM | Acute/subacute | Anti-MDA5 antibody (22) |
| Ikeda2015 [32] | Japan | 16 | Retrospective cohort | 689 days (median) | CADM | Not specified | Anti-MDA5 antibody (10)<br>Anti-ARS antibody (3)[e] |
| Hozumi2015 [21] | Japan | 48 | Retrospective cohort | 0.2-19.2 years (range) | PM, DM, CADM | Acute/subacute, Chronic | Anti-ARS antibody (23)[f] |
| Rojas-Serrano2015 [33] | Mexico | 43 | Retrospective cohort | Not specified | ASS | Not specified | Anti-ARS antibody (43)[g] |
| Takada2015 [34] | Japan | 13 | Case-control | Not specified | ADM | Not specified | Not specified |
| Zou2015 [26] | China | 37 | Retrospective cohort | Not specified | CADM | Acute | Anti-MDA5 antibody (34) |
| Fujisawa2014 [22] | Japan | 114 | Retrospective cohort | Not specified | PM, DM, CADM | Acute/subacute, Chronic | Anti-Jo-1 antibody (19.3%) |
| Nara2014 [35] | Japan | 12 | Case-control | Not specified | CADM | Rapidly progressive | Anti-MDA5 antibody (11) |

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| Chen2013 [36] | China | 14 | Case-control | Not specified | DM | Acute | Anti-Jo-1 antibody (2) |
| Muro2013 [37] | Japan | 25 | Case-control | Not specified | DM, CADM | Rapidly progressive, Chronic | Anti-MDA5 antibody (25) |
| Sato2013 [38] | Japan | 10 | Case-control | Not specified | DM, CADM | Rapidly progressive | Anti-MDA5 antibody (10) |
| Shimojima2013 [39] | Japan | 21 | Case-control | Not specified | DM, CADM | Active | Anti-Jo-1 antibody (1) |
| Sun2013 [40] | Japan | 25 | Retrospective cohort | Not specified | CADM | Acute/subacute, Chronic | Anti-Jo-1 antibody (1) |
| Tanizawa2013 [41] | Japan | 51 | Retrospective cohort | 714 (26-2418) days | PM, DM, CADM | Not specified | Anti-MDA5 antibody (21) |
| | | | | (median (range)) | | | Anti-ARS antibody (20)[h] |
| Gono2012 [23] | Japan | 20 | Case-control | Not specified | DM, CADM | Rapidly progressive | Anti-MDA5 antibody (20) |
| Yamasaki2011 [42] | Japan | 69 | Retrospective cohort | Not specified | DM, CADM | Not specified | Not specified |
| Gono-Satoh2010 [24] | Japan | 14 | Retrospective cohort | Not specified | DM, CADM | Acute/subacute, Chronic | Anti-MDA5 antibody (14) |
| Su-yun2010 [43] | China | 69 | Retrospective cohort | Not specified | PM, DM, ADM | Hamman-Rich-like | Anti-Jo-1 antibody (5) |
| | | | | | | Slowly progressive | |
| | | | | | | Asymptomatic | |
| Mukae2009 [44] | Japan | 27 | Retrospective cohort | Not specified | DM, CADM | Acute, Chronic | Anti-Jo-1 antibody (5) |
| Hayashi2008 [45] | Japan | 33 | Retrospective cohort | Not specified | PM, DM | Not specified | Not specified |
| Huh2007 [46] | Korea | 33 | Retrospective cohort | Not specified | PM, DM | Acute, Chronic | Anti-Jo-1 antibody (4) |
| Ye2007 [27] | China | 49 | Retrospective cohort | 1-63 months (range) | PM. DM | Not specified | Anti-ARS antibody (6)[i] |
| Kameda2005 [47] | Japan | 10 | Case-control | Not specified | DM | Acute/subacute | Not specified |
| Kang2005 [48] | Korea | 29 | Retrospective cohort | Not specified | PM, DM, ADM | Hamman-Rich-like | Anti-Jo-1 antibody (6) |
| | | | | | | Slowly progressive | |
| | | | | | | Asymptomatic | |
| Ito1999 [49] | Japan | 16 | Case-control | Not specified | DM | Rapidly progressive, Chronic | Anti-Jo-1 antibody (3) |
| Tazelaar1990 [50] | USA | 15 | Case-control | Not specified | PM, DM | Not specified | Not specified |

a, Only 4 studies (Kurasama2018, Sato2013, Yamasaki2011, Mukae2009) provided the ethnicity of participants, all of which were Japanese except one non-Japanese Asian patient who participated in Yamasaki2011.

b, Only data of anti-ARS antibody and anti-MDA5 antibody were retrieved.

c, including 21 cases of anti-Jo-1 antibody

d, Anti-PL-7 antibody (2), anti-EJ antibody (1)

e, Anti-Jo-1 antibody (1), Anti-PL12 antibody (1), Anti-OJ antibody (1)

f, Anti-Jo-1 antibody (6), Anti-PL7 antibody (8), Anti-PL12 antibody (4), Anti-KS antibody (2), Anti-EJ antibody (2), Anti-KS antibody plus anti-EJ antibody (1)

g, Anti-Jo-1 antibody (35), Anti-PL7 antibody (2), Anti-EJ antibody (6)

h, Anti-Jo-1 antibody (9), Anti-PL7 antibody (4), Anti-PL12 antibody (1), Anti-EJ antibody (3), Anti-OJ antibody (2), unspecified (1)

i, Anti-Jo-1 antibody (5), Anti-PL7 antibody (1)

ADM, amyopathic dermatomyositis; anti-ARS antibody, anti-aminoacyl-transfer RNA synthetase antibody; anti-MDA5 antibody, anti-melanoma differentiation antigen 5 antibody; ASS, anti-synthetase syndrome; CADM, clinically amyopathic dermatomyositis; DM, dermatmyositis; ILD, interstitial lung disease; PM, polymyositis;

**e-Table 3 Risk of bias in 32 studies reporting potential prognostic factors for all-cause mortality assessed by the Quality in Prognostic Studies tool[a]**

| Study | study participation | study attrition[b] | prognostic factor measurement | outcome measurement | study confounding | statistical analysis and reporting |
|---|---|---|---|---|---|---|
| Hozumi2018 [19] | low risk | **high risk** | low risk | low risk | **high risk** | **high risk** |
| Kurasawa2018 [28] | medium risk | low risk | **high risk** | low risk | **high risk** | **high risk** |
| Sugiyama2018 [29] | **high risk** | low risk | medium risk | low risk | **high risk** | **high risk** |
| Zhang2018 [30] | low risk | **high risk** | **high risk** | low risk | **high risk** | **high risk** |
| Enomoto2017 [20] | low risk | **high risk** | low risk | low risk | **high risk** | **high risk** |
| Okabayashi2017 [31] | **high risk** | low risk | medium risk | low risk | **high risk** | **high risk** |
| Lin2016 [25] | low risk | **high risk** | **high risk** | low risk | **high risk** | **high risk** |
| Ikeda2015 [32] | low risk | **high risk** | low risk | low risk | **high risk** | **high risk** |
| Hozumi2015 [21] | low risk | **high risk** | low risk | low risk | **high risk** | **high risk** |
| Rojas-Serrano2015 [33] | **high risk** | low risk | low risk | low risk | **high risk** | **high risk** |
| Takada2015 [34] | **high risk** | N/A | low risk | low risk | **high risk** | **high risk** |
| Zou2015 [26] | **high risk** | low risk | **high risk** | low risk | **high risk** | **high risk** |
| Fujisawa2014 [22] | low risk | **high risk** | low risk | low risk | **high risk** | **high risk** |
| Nara2014 [35] | **high risk** | N/A | low risk | low risk | **high risk** | **high risk** |
| Chen2013 [36] | **high risk** | N/A | **high risk** | medium risk | **high risk** | **high risk** |
| Muro2013 [37] | low risk | N/A | low risk | low risk | **high risk** | **high risk** |
| Sato2013 [38] | **high risk** | N/A | low risk | low risk | **high risk** | **high risk** |
| Shimojima2013 [39] | **high risk** | N/A | medium risk | low risk | **high risk** | **high risk** |

| | | | | | | |
|---|---|---|---|---|---|---|
| Sun2013 [40] | **high risk** | **high risk** | low risk | low risk | **high risk** | **high risk** |
| Tanizawa2013 [41] | **high risk** | **high risk** | low risk | low risk | **high risk** | **high risk** |
| Gono2012 [23] | **high risk** | N/A | low risk | low risk | **high risk** | **high risk** |
| Yamasaki2011 [42] | **high risk** | **high risk** | low risk | low risk | **high risk** | **high risk** |
| Gono-Satoh2010 [24] | **high risk** | **high risk** | low risk | low risk | **high risk** | **high risk** |
| Su-yun2010 [43] | low risk | **high risk** | **high risk** | low risk | **high risk** | **high risk** |
| Mukae2009 [44] | low risk | low risk | **high risk** | low risk | **high risk** | **high risk** |
| Hayashi2008 [45] | **high risk** | **high risk** | medium risk | low risk | **high risk** | **high risk** |
| Huh2007 [46] | low risk | **high risk** | low risk | low risk | **high risk** | **high risk** |
| Ye2007 [27] | low risk | **high risk** | low risk | low risk | **high risk** | **high risk** |
| Kameda2005 [47] | medium risk | N/A | **high risk** | low risk | **high risk** | **high risk** |
| Kang2005 [48] | medium risk | **high risk** | **high risk** | low risk | **high risk** | **high risk** |
| Ito1999 [49] | **high risk** | N/A | **high risk** | low risk | **high risk** | **high risk** |
| Tazelaar1990 [50] | **high risk** | N/A | low risk | low risk | **high risk** | **high risk** |

a, Text in bold referring to high risk of bias;

b, N/A indicating not applicable due to case-control studies;

## e-Table 4 The result of univariate analysis of potential prognostic factors for all-cause mortality

| Potential prognostic factors[a] | Analysis | Studies[b] (n) | Participants (n) | +[c] | -[d] | Significance (n) | Result of meta-analysis and non-pooled studies[e] (95% confidence interval unless otherwise specified) |
|---|---|---|---|---|---|---|---|
| Demographic features | | | | | | | |
| ***Age*** | Meta | 9 | 267 | 7 | 1 | 1 | **MD 5.90 (3.17-8.63) (years)** |
| | Not pooled | Zou2015 [26] | 37 | 1 | 0 | 0 | HR 1.01 (0.95-1.05) (/1year) |
| | | Fujisawa2014 [22] | 114 | 1 | 0 | 1 | **HR 1.05 (1.01-1.08) (/1year)** |
| | | Ikeda2015 [32] | 10 | 1 | 0 | 0 | Median (IQR) 63.0 (59.3-64.5)/58.5 (47.8-64.3) (dead/alive) (years) |
| | | Okabayashi2017[31] | 14 | 0 | 1 | 0 | Median 58/64 (dead/alive) (years) |
| | | Zhang2018[30] | 103 | 1 | 0 | 1 | **RR 2.09 (1.28-3.42) (>60 vs <60 years )** |
| Sex (Female) | Meta | 16 | 586 | 5 | 11 | 0 | OR 0.85 (0.63-1.15) (vs male) |
| CADM | Meta | 7 | 320 | 3 | 4 | 0 | OR 1.32 (0.85-2.06) (vs DM) |
| ***Dermatomyositis*** | Meta | 5 | 290 | 5 | 0 | 2 | **OR 2.73 (1.89-3.94) (vs PM)** |
| ***Types of ILD (A/SIP)*** | Meta | 10 | 460 | 10 | 0 | 6 | **OR 4.85 (2.81-8.37) (vs chronic IP)** |
| Symptoms | | | | | | | |
| ***Fever*** | Meta | 8 | 391 | 6 | 2 | 2 | **OR 1.81 (1.20-2.74)** |
| Cough | Meta | 5 | 194 | 1 | 4 | 0 | OR 0.75 (0.40-1.38) |
| Dyspnea | Meta | 5[f] | 194 | 3 | 1 | 0 | OR 1.42 (0.85-2.38) |
| | Not pooled | Tanizawa2013 [41] | 51 | 1 | 0 | 0 | HR 1.06 (0.14-6.32) (British Medical Research Council dyspnea scale) |
| Muscle weakness | Meta | 5 | 228 | 4 | 1 | 0 | OR 1.60 (0.81-3.15) |
| | Not pooled | Ito1999 [49] | 9 | 0 | 1 | 1 | **MD 1.10 (0.08-2.13) (the manual muscle test)** |
| Pulmonary function tests | | | | | | | |

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| ***%FVC*** | Meta | 4 | 245 | 0 | 4 | 2 | **OR 0.96 (0.95-0.98) (/1%)** |
| %DLCO | Meta | 3 | 121 | 0 | 3 | 0 | **OR 0.97 (0.94-0.99) (/1%)** |
| | Not pooled | Shimojima2013 [39] | 21 | 0 | 1 | 1 | **MD -15.3 (-25.3- -5.3) (%)** |
| | | Gono2012 [23] | 7 | 1 | 0 | -[g] | Median (IQR) 10 (8.9-12.7)/9.5 (7.8-13.2) (dead/alive) (ml/min/mmHg) |
| Radiological findings on HRCT | | | | | | | |
| GGO/GGA | Meta | 5 | 202 | 5 | 0 | 0 | OR 1.51 (0.88-2.60) |
| | Not pooled | Rojas2015 [33] | 43 | 1 | 0 | 1 | **HR 5.87 (1.43-24.01) (Kazerooni groud-glass score)** |
| Consolidation | Meta | 4 | 151 | 1 | 3 | 0 | OR 0.78 (0.48-1.26) |
| **Reticular opacity** | Not pooled | Rojas2015 [33] | 43 | 1 | 0 | 0 | HR 1.06 (0.92-1.23) (Goh score) |
| | | Tanizawa2013 [41] | 51 | 1 | 0 | 0 | HR 1.04 (0.97-1.10) (Goh score) |
| | | Hayashi2008 [45] | 16 | 1 | 0 | 0 | RR 2.27 (0.35-14.61) |
| | | Ikeda2015 [32] | 10 | 0 | 1 | 0 | RR 0.38 (0.03-4.44) |
| Extent of abnormality | Not pooled | Zou2015 [26] | 37 | 1 | 0 | 1 | **HR 1.15 (1.05-1.27) (/5)** |
| | | Tanizawa2013 [41] | 51 | 0 | 1 | 0 | HR 0.99 (0.95-1.02) (Goh score) |
| | | Shimojima2013[39] | 21 | 1 | 0 | 1 | **MD 8.49 (2.72-14.26)** |
| | | Okabayashi2017[31] | 14 | 0 | 1 | 0 | Median 231.4/247.8 (dead/alive) (%) |
| Laboratory findings | | | | | | | |
| ***Anti-Jo-1 antibody*** | Meta | 6 | 403 | 0 | 6 | 1 | **OR 0.35 (0.18-0.71)** |
| ***Anti-MDA5 antibody*** | Meta | 5 | 165 | 5 | 0 | 3 | **OR 3.35 (1.55-7.25)** |
| | Not pooled | Li2016[25] | 26 | 1 | 0 | 0 | Positivity 90.9/80.0 (dead/alive) (%) |
| Anti-MDA5 antibody titer | Meta | 4 | 51 | 3 | 1 | 2 | SMD 0.89 (-0.14-1.92) |
| | Not pooled | Zou2015 [26] | 37 | - | - | 0 | HR 1.00 (0.998-1.002) (/1U/mL) |

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| | | Gono2012 [23] | 20 | 1 | 0 | 0 | Median (IQR) 332.1 (92.0-599.8)/129.3 (44.6-254.0) (dead/alive) (U/mL) |
| ANA | Meta | 6 | 295 | 4 | 2 | 0 | OR 0.89 (0.55-1.44) |
| CK | Meta | 6 | 79 | 4 | 2 | 1 | SMD 0.15 (-0.41-0.71) |
| | Not pooled | Rojas2015 [33] | 43 | 0 | 1 | 0 | HR 0.9998 (0.9993-1.0002) (/1U/L) |
| | | Fujisawa2014 [22] | 114 | 0 | 1 | 0 | HR 0.9997 (0.9993-1.0000) (/1IU/L) |
| | | Yamasaki2011 [42] | 69 | 1 | 0 | 1 | **HR 2.51 (1.87-3.37) (<500IU/L)** |
| | | Gono2012 [23] | 20 | 0 | 1 | 0 | Median (IQR) 77 (62-324)/95 (38-383) (dead/alive) (IU/L) |
| | | Sugiyama2018[29] | 116 | 1 | 0 | 0 | Median (IQR) 377 (118-1083)/301 (118-1691) (dead/alive) (U/L) |
| | | Okabayashi2017[31] | 14 | 0 | 1 | 0 | Median 65/69 (dead/alive) (U/L) |
| | | Zhang2018[30] | 103 | 1 | 0 | 0 | RR 1.27 (0.75-2.17) (>194 IU/L vs normal) |
| Aldolase | Not pooled | Nara2014 [35] | 12 | 1 | 0 | 0 | MD 1.40 (-3.57-6.37) (IU/L) |
| | | Huh2007 [46] | 11 | 0 | 1 | 0 | MD -9.90 (-65.61-45.81) (SU/mL) |
| | | Fujisawa2014 [22] | 114 | 0 | 1 | 0 | HR 0.99 (0.97-1.01) (/1IU/L) |
| KL-6 | Meta | 3 | 43 | 2 | 1 | 0 | SMD 0.24 (-0.37-0.84) |
| | Not pooled | Fujisawa2014 [22] | 114 | 0 | 1 | 0 | HR 0.9999 (0.9994-1.0003) (/1U/mL) |
| | | Ikeda2015 [32] | 10 | 0 | 1 | 0 | Median (IQR) 524 (470-626)/1135 (930-1190) (dead/alive) (U/mL) |
| | | Gono2012 [23] | 18 | 0 | 1 | 0 | Median (IQR) 731 (602-1099)/842 (678-1009) (dead/alive) (U/mL) |
| | | Okabayashi2017[31] | 14 | 1 | 0 | 0 | Median 2024/906 (dead/alive) (U/mL) |
| LDH | Meta | 4 | 52 | 3 | 1 | 1 | SMD 0.64 (-0.28-1.56) |
| | Not pooled | Zou2015 [26] | 37 | 1 | 0 | 1 | **HR 1.005 (1.003-1.008) (/1U/L)** |
| | | Fujisawa2014 [22] | 114 | 0 | 1 | 0 | HR 0.9999 (0.9992-1.0000) (/1IU/L) |
| | | Gono2012 [23] | 20 | 1 | 0 | 0 | Median (IQR) 460 (308-518)/364(243-488) (dead/alive) (IU/L) |
| | | Sugiyama2018[29] | 116 | 1 | 0 | 0 | Median (IQR) 438 (337-538)/364(276-516) (dead/alive) (U/L) |

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| | | Okabayashi2017[31] | 14 | 1 | 0 | 1 | **Median 468/332 (dead/alive) (U/L)** |
| | | Zhang2018[30] | 103 | 1 | 0 | 0 | RR 2.90 (0.78-10.81) (>215 U/L vs normal) |
| ***Ferritin*** | Not pooled | Zou2015 [26] | 37 | 1 | 0 | 1 | **HR 1.001 (1.000-1.002) (/1ng/mL)** |
| | | Hozumi2018[19] | 69 | 1 | 0 | 1 | **HR 1.03 (1.01-1.04) (/100ng/mL)** |
| | | Tanizawa2013 [41] | 51 | 1 | 0 | 1 | **HR 3.22 (1.16-8.47) (>500ng/mL)** |
| | | Kurasawa2018[28] | 15 | 1 | 0 | 1 | **log rank (p=0.01) (>1000 vs <=1000) (ng/mL)** |
| | | Nara2014 [35] | 12 | 1 | 0 | 0 | MD 329.3 (-293.1-951.7) (ng/mL) |
| | | Muro2013 [37] | 17 | 1 | 0 | 1 | **MD 1690 (224-3156) (ng/mL)** |
| | | Gono2012 [23] | 20 | 1 | 0 | 1 | **Median (IQR) 1600 (835-1935)/409 (248-843) (dead/alive) (ng/mL)** |
| | | Sugiyama2018[29] | 116 | 1 | 0 | 0 | Median (IQR) 658 (148-1210)/342 (165-734) (dead/alive) (ng/mL) |
| | | Okabayashi2017[31] | 14 | 1 | 0 | 1 | **Median 1260.3/180.6 (dead/alive) (ng/mL)** |
| ***A-aDO2*** | Not pooled | Nara2014 [35] | 12 | 1 | 0 | 0 | MD 28.3 (-23.5-80.1) (mmHg) |
| | | Chen2013 [36] | 14 | 1 | 0 | 1 | **MD 352.9 (192.2-513.6)** |
| | | Gono2012 [23] | 20 | 1 | 0 | 1 | **Median (IQR) 41 (30-102)/26 (22-34) (dead/alive) (mmHg)** |
| ***PaO2*** | Meta | 4 | 66 | 0 | 4 | 3 | **MD -12.35 (-22.45- -2.25) (mmHg)** |
| | Not pooled | Hozumi2018[19] | 69 | 0 | 1 | 1 | **HR 0.49 (0.33-0.74) (/10 mmHg)** |
| **CRP** | Not pooled | Zou2015 [26] | 37 | 1 | 0 | 0 | HR 1.02 (0.85-1.23) (/1mg/L) |
| | | Fujisawa2014 [22] | 114 | 1 | 0 | 0 | HR 1.11 (1.00-1.21) (/1mg/dL) |
| | | Chen2013 [36] | 14 | 1 | 0 | 0 | MD 36.31 (-1.84-74.46) (mg/L) |
| | | Nara2014 [35] | 12 | 1 | 0 | 0 | MD 2.08 (-3.21-7.37) (mg/dL) |
| | | Gono2012 [23] | 20 | 1 | 0 | 0 | Median (IQR) 1.06 (0.17-2.16)/0.63 (0.10-1.96) (dead/alive) (mg/dL) |
| | | Sugiyama2018[29] | 116 | 1 | 0 | 0 | Median (IQR) 1.32 (0.58-1.96)/0.49 (0.12-1.62) (dead/alive) (mg/dL) |
| | | Okabayashi2017[31] | 14 | 0 | 1 | 0 | Median 0.56/4.08 (dead/alive) (mg/dL) |

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| | | Zhang2018[30] | 103 | 1 | 0 | 1 | **RR 2.12 (1.19-3.77) (>10mg/L vs normal)** |
| ***AST*** | Not pooled | Nara2014 [35] | 12 | 1 | 0 | 0 | MD 67.8 (-80.3-215.9) (IU/L) |
| | | Chen2013 [36] | 14 | 1 | 0 | 0 | MD 37.8 (-17.4-92.9) (U/L) |
| | | Ikeda2015 [32] | 10 | - | - | 0 | Median (IQR) 55.5 (41.0-110)/55.5 (36.8-76.8) (dead/alive) (IU/L) |
| | | Gono-Satoh2010 [24] | 14 | 1 | 0 | 1 | **Median (IQR) 175 (110-421/70 (35-142) (dead/alive) (IU/L)** |
| | | Zhang2018[30] | 103 | 1 | 0 | 0 | RR 1.22 (0.72-2.08) (>40U/L vs normal) |

a, Text in italic bold referring to potential prognostic factors, which demonstrated that the summary effect was statistically significant by meta-analysis and where data were not combined, the effect was consistent in the majority of the included studies (≥75%);

b, the number indicating included studies for meta-analysis;

c, indicating a positive association between all-cause mortality and potential prognostic factors based on a point estimate;

d, indicating a negative association between all-cause mortality and potential prognostic factors based on a point estimate;

e, Text in bold referring to statistically significant results;

f, one study showed a null value for the association between all-cause mortality and dyspnea based on a point estimate;

g, no statistical analysis was undertaken;

A-aDO2, alveolar-arterial oxygen difference; ANA, anti-nuclear antibody; anti-MDA5 antibody, anti-melanoma differentiation antigen 5 antibody; A/SIP, acute/subacute interstitial pneumonia; CADM, clinically amyopathic dermatomyositis; CK, creatine kinase; CRP, C-reactive protein; %DLCO, percentage of predicted diffusion capacity of the lung for carbon monoxide; DM, dermatomyositis; %FVC, percentage of predicted forced vital capacity; GGA, ground glass attenuation; GGO, ground glass opacity; HR, hazard ratio; HRCT, high resolution computed tomography; IP, interstitial pneumonia; IQR, interquartile range; KL-6, Krebs von

den Lungen-6; LDH, lactate dehydrogenase; MD, mean difference; Meta, meta-analysis; SMD, standardized mean difference; OR, odds ratio; PaO2, arterial oxygen pressure; PM, polymyositis; RR, risk ratio;

## e-Table 5 The result of multivariate analysis of potential prognostic factors for all-cause mortality

| Potential prognostic factors[a] | Analysis | Studies[b] (n) | Participants (n) | +[c] | -[d] | Significance (n) | Result of meta-analysis and non-pooled studies[e] (95% confidence interval unless otherwise specified) |
|---|---|---|---|---|---|---|---|
| Demographic features | | | | | | | |
| ***Age*** | Not pooled | Fujisawa2014 [22] | 114 | 1 | 0 | 1 | **HR 1.06 (1.02-1.10) (/1year)** |
| | | Zhang2018[30] | 103 | 1 | 0 | 1 | **HR 2.31 (1.06-5.06) (>60 vs <60 years )** |
| CADM | Not pooled | Fujisawa2014 [22] | 84 | 1 | 0 | 0 | HR 1.91 (0.79-4.90) (vs DM) |
| Dermatomyositis | No pooled | Fujisawa2014 [22] | 71 | 1 | 0 | 0 | HR 2.19 (0.62-10.10) (vs PM) |
| ***Types of ILD (A/SIP)*** | Not pooled | Fujisawa2014 [22] | 114 | 1 | 0 | 1 | **HR 4.23 (1.69-12.09) (vs chronic IP)** |
| | | Zhang2018[30] | 103 | 1 | 0 | 1 | **HR 5.17 (1.94-13.49) (vs chronic IP)** |
| Symptoms | | | | | | | |
| Fever | Not pooled | Tanizawa2013 [41] | 51 | 1 | 0 | 0 | HR 1.59 (0.51-5.03) |
| | | Enomoto2017[20] | 48 | 1 | 0 | 1 | **HR 4.68 (1.36-16.10)** |
| Cough | Not pooled | Enomoto2017[20] | 48 | 0 | 1 | 0 | HR 0.76 (0.22-2.67) |
| Dyspnea | Not pooled | Zhang2018[30] | 103 | 1 | 0 | 0 | HR 1.40 (0.63-3.10) |
| | | Enomoto2017[20] | 48 | 1 | 0 | 0 | HR 2.65 (0.78-9.03) |
| Muscle weakness | Not pooled | Enomoto2017[20] | 48 | 1 | 0 | 0 | HR 2.44 (0.63-9.48) |
| Pulmonary function tests | | | | | | | |
| ***%FVC*** | Not pooled | Fujisawa2014 [22] | 114 | 0 | 1 | 1 | **HR 0.96 (0.93-0.99) (/1%)** |
| Radiological findings on HRCT | | | | | | | |
| ***GGO/GGA*** | Not pooled | Rojas2015 [33] | 43 | 1 | 0 | 1 | **HR 7.68 (1.37-43.03) (Kazerooni ground-glass score)** |
| ***Extent of abnormality*** | Not pooled | Zou2015 [26] | 37 | 1 | 0 | 1 | **HR 1.13 (1.01-1.28) (/5)** |

| | | | | | | | |
|---|---|---|---|---|---|---|---|
| ***(HRCT)*** | | | | | | | |
| Laboratory findings | | | | | | | |
| ***Anti-Jo-1 antibody*** | Not pooled | Rojas2015 [33] | 43 | 0 | 1 | 1 | **HR 0.004 (0.00003-0.54)** |
| Anti-MDA5 antibody | Not pooled | Tanizawa2013 [41] | 51 | 1 | 0 | 1 | **HR 4.97 (1.55-19.30)** |
| | | Enomoto2017[20] | 48 | 1 | 0 | 0 | HR 3.11 (0.90-10.74) |
| CK | Not pooled | Nara2014 [35] | 12 | 1 | 0 | 0 | OR 1.02 (0.99-1.05) (/1IU/L) |
| | | Enomoto2017[20] | 48 | - | - | 0 | HR 1.00 (0.99-1.01) (/10IU/mL) |
| Aldolase | Not pooled | Nara2014 [35] | 12 | 0 | 1 | 0 | OR 0.64 (0.35-1.14) (/1IU/L) |
| KL-6 | Not pooled | Nara2014 [35] | 12 | - | - | 0 | OR 1.00 (0.999-1.002) (/1U/mL) |
| | | Enomoto2017[20] | 48 | 1 | 0 | 0 | HR 1.05 (1.00-1.12) (/100U/mL) |
| LDH | Not pooled | Zou2015 [26] | 37 | 1 | 0 | 0 | HR 1.004 (1.000-1.009) (/1U/L) |
| | | Zhang2018[30] | 103 | 1 | 0 | 0 | RR 2.56 (0.48-13.73) (>215 U/L vs normal) |
| Ferritin | Meta | 4 | 132 | 3 | 1 | 2 | HR 1.00 (1.00-1.01) (/10ng/mL) |
| | Not pooled | Tanizawa2013 [41] | 51 | 1 | 0 | 0 | HR 1.37 (0.44-4.17) (>500ng/mL) |
| A-aDO2 | Not pooled | Gono-Satoh2010 [24] | 14 | - | - | 0 | HR 1.00 (0.99-1.03) (/1mmHg) |
| PaO2 | Not pooled | Hozumi2018[19] | 69 | 0 | 1 | 0 | HR 0.62 (0.32-1.18) (/10mmHg) |
| CRP | Not pooled | Nara2014 [35] | 12 | 1 | 0 | 0 | OR 1.17 (0.21-6.33) (/1mg/dL) |
| | | Enomoto2017[20] | 48 | 1 | 0 | 0 | HR 1.13 (0.91-1.41) (/1mg/dL) |
| | | Zhang2018[30] | 103 | 1 | 0 | 1 | **HR 2.74 (1.18-6.37) (>10mg/L vs normal)** |
| AST | Not pooled | Gono-Satoh2010 [24] | 14 | 1 | 0 | 0 | HR 1.005 (1.00-1.01) (/1IU/L) |

a, Text in italic bold referring to items determined as prognostic factors, which demonstrated that the summary effect was statistically significant by meta-analysis and where data were not combined, the effect was consistent and significant in the majority of the included studies (≥75%);

b, the number indicating included studies for meta-analysis;

c, indicating a positive association between all-cause mortality and potential prognostic factors based on a point estimate;

d, indicating a negative association between all-cause mortality and potential prognostic factors based on a point estimate;

e, Text in bold referring to statistically significant results;

A-aDO2, alveolar-arterial oxygen difference; anti-MDA5 antibody, anti-melanoma differentiation antigen 5 antibody; A/SIP, acute/subacute interstitial pneumonia; CADM, clinically amyopathic dermatomyositis; CK, creatine kinase; CRP, C-reactive protein; %DLCO, percentage of predicted diffusion capacity of the lung for carbon monoxide; DM, dermatomyositis; %FVC, percentage of predicted forced vital capacity; GGA, ground glass attenuation; GGO, ground glass opacity; HR, hazard ratio; HRCT, high resolution computed tomography; IP, interstitial pneumonia; KL-6, Krebs von den Lungen-6; LDH, lactate dehydrogenase; Meta, meta-analysis; OR, odds ratio; PaO2, arterial oxygen pressure; PM, polymyositis;

e-**Table 6 Assessment of quality of evidence of prognostic factors by the Grades of Recommendation, Assessment, Development and Evaluation (GRADE) system**

| **Outcome: all-cause mortality** | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|
| | | | GRADE factors | | | | | | | |
| Prognostic factors[a] | Analysis[b] | Phase | Study limitations | Inconsistency[c] | Indirectness | Publication bias | Imprecision | Moderate/large effect size | Dose effect | Overall quality |
| Age | Uni | 1 | + | - | - | + | - | + | - | Low |
| | Multi | 1 | + | - | - | + | + | - | - | Very low |
| Types of ILD (A/SIP) | Uni | 1 | + | - | - | + | - | + | - | Low |
| | Multi | 1 | + | - | - | + | + | + | - | Very low |
| %FVC | Uni | 1 | + | - | - | + | - | - | - | Very low |
| | Multi | 1 | + | N/A | - | + | + | - | - | Very low |
| GGO/GGA | Uni | 1 | + | - | - | + | - | - | - | Very low |
| | Multi | 1 | + | N/A | + | + | + | + | - | Very low |
| Extent of abnormality | Uni | 1 | + | + | - | + | + | - | - | Very low |
| (HRCT) | Multi | 1 | + | N/A | + | + | + | - | - | Very low |
| Anti-Jo-1 antibody | Uni | 1 | + | - | + | + | - | - | - | Very low |
| | Multi | 1 | + | N/A | + | + | + | + | - | Very low |

a, 6 items were determined as prognostic factors from a total of 28 potential prognostic factors based on the significant result of meta-analysis and the consistent and significant result of non-pooled studies on multivariate analysis.

b, ‘uni’ indicating univariate analysis while ‘multi’ indicating multivariate analysis.

c, N/A indicating not applicable due to only one study available.

A/SIP, acute/subacute interstitial pneumonia; %FVC, percentage of predicted forced vital capacity; GGA, ground glass attenuation; GGO, ground glass opacity; HRCT, high resolution computed tomography;

e-Appendix: Search terms for each electronic database

Ovid Medline (through the 9th of August, 2018)

1 exp Polymyositis/

2 exp Dermatomyositis/

3 exp Myositis/

4 polymyositis.mp.

5 dermatomyositis.mp.

6 myositis.mp.

7 myopath$.mp.

8 PM.mp.

9 DM.mp.

10 (anti$synthetase adj syndrome).mp.

11 exp Lung Diseases, Interstitial/

12 exp Pulmonary Fibrosis/

13 (interstitial adj3 lung adj3 disease$).mp.

14 (interstitial adj3 pneumoni$).mp.

15 (interstitial adj3 pneumopath$).mp.

16 alveolitis.mp.

17 (pulmonary adj3 fibros$).mp.

18 incidence.sh.

19 exp Mortality/

20 follow-up studies.sh.

21 prognos$.tw.

22 predict$.tw.

23 course$.tw.

24 1 or 2 or 3 or 4 or 5 or 6 or 7 or 8 or 9 or 10

25 11 or 12 or 13 or 14 or 15 or 16 or 17

26 18 or 19 or 20 or 21 or 22 or 23

27 24 and 25 and 26

Ovid EMBASE (through the 9th of August, 2018)

1 exp polymyositis/

2 exp dermatomyositis/

3 exp myositis/

4 polymyositis.mp.

5 dermatomyositis.mp.

6 myositis.mp.

7 myopath$.mp.

8 PM.mp.

9 DM.mp.

10 (anti$synthetase adj syndrome).mp.

11 exp interstitial lung disease/

12 exp lung fibrosis/

13 (interstitial adj3 lung adj3 disease$).mp.

14 (interstitial adj3 pneumoni$).mp.

15 (interstitial adj3 pneumopath$).mp.

16 alveolitis.mp.

17 (pulmonary adj3 fibros$).mp.

18 exp disease course/

19 risk$.mp.

20 diagnos$.mp.

21 follow-up.mp.

22 ep.fs.

23 outcome.tw.

24 1 or 2 or 3 or 4 or 5 or 6 or 7 or 8 or 9 or 10

25 11 or 12 or 13 or 14 or 15 or 16 or 17

26 18 or 19 or 20 or 21 or 22 or 23

27 24 and 25 and 26

Science Citation Index Expanded (Web of Science) (through the 9th of August, 2018)

#1 TS=(polymyositis) OR TS=(dermatomyositis) OR TS=(myositis) OR TS=

(myopath*) OR TS=("anti$synthetase syndrome")

#2 TS=("interstitial NEAR/3 disease$") OR TS=("interstitial NEAR/3 pneumoni*") OR TS=("interstitial NEAR/3 pneumopath*") OR TS=(alveolitis) OR TS=("pulmonary NEAR/3 fibros*")

#3 TS=(prognos*) OR TS=(mortality) OR TS=(outcome) OR TS=(course$) OR TS=(follow-up) OR TS=(predict*) OR TS=(incidence) OR TS=(risk)

#4 #3 AND #2 AND #1

Google Scholar

(polymyositis OR dermatomyositis OR myositis OR "antisynthetase syndrome") (“interstitial lung disease” OR “interstitial pneumonia” OR “interstitial pneumopathy” OR “pulmonary fibrosis”) (prognosis OR mortality OR outcome)