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1 **The accuracy of Japanese administrative data in identifying acute exacerbation of**
2 **idiopathic pulmonary fibrosis**

3 **Running title: Japanese administrative data in AE-IPF identification**

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34 **Keywords: Idiopathic pulmonary fibrosis, Acute exacerbation, Validation,**

35 **Administrative data**

36 **Abstract**

37 **Background:** This study aimed to develop criteria for identifying patients with acute
38 exacerbation of idiopathic pulmonary fibrosis (AE-IPF) from Japanese administrative
39 data and validate the pre-existing criteria.

40 **Methods:** This retrospective, multi-center validation study was conducted at eight
41 institutes in Japan to verify the diagnostic accuracy of the disease name for AE-IPF. We
42 used the Japanese Diagnosis Procedure Combination data to identify patients with a
43 disease name that could meet the diagnostic criteria for AE-IPF, who were admitted to
44 the eight institutes from January 2016 to February 2019. As a reference standard, two
45 respiratory physicians performed a chart review to determine whether the patients had a
46 disease that met the diagnostic criteria for AE-IPF. Furthermore, two radiologists
47 interpreted the chest computed tomography findings of cases considered AE-IPF and
48 confirmed the diagnosis. We calculated the positive predictive value (PPV) for each
49 disease name and its combination.

50 **Results:** We included 830 patients; among them, 216 were diagnosed with AE-IPF
51 through the chart review. We combined the groups of disease names and yielded two
52 criteria: the criteria with a high PPV (0.72 [95% confidence interval 0.62 to 0.81]) and

53 that with a slightly less PPV (0.61 [0.53 to 0.68]) but more true positives. Pre-existing
54 criteria showed a PPV of 0.40 (0.31 to 0.49).

55 **Conclusion:** The criteria derived in this study for identifying AE-IPF from Japanese
56 administrative data show a fair PPV. Although these criteria should be carefully
57 interpreted according to the target population, our findings could be utilized in future
58 database studies on AE-IPF.

59 **Background**

60 Idiopathic pulmonary fibrosis (IPF) is a rare disease with a poor prognosis that is
61 characterized by chronic progressive lung fibrosis. Although IPF is rare, it has a worse
62 prognosis than numerous cancer types.[1] Moreover, hospitalization and mortality rates
63 attributable to IPF have increased over recent years, which is suggestive of an
64 increasing burden of this disease on healthcare services worldwide.[2-4] During the
65 disease course, acute exacerbation (AE) may occur at an annual frequency of
66 approximately 5–15%; further, the mortality rate is reported to be approximately 50–
67 60%.[5, 6] Few randomized controlled trials (RCTs) with limited external validity have
68 investigated AE-IPF.[7, 8] Therefore, there is a need for large observational studies on a
69 wider patient population using real-world data to complement RCTs.[9]

70 However, there have been rare seen large-scale database studies on AE-IPF.
71 This could be attributed to the difficulty in identifying cases. In large-scale database
72 research, case identification is essential, with validation studies being crucial for this
73 purpose. The REporting of studies Conducted using Observational Routinely collected
74 health Data (RECORD) statement mentions that “Any validation studies of the codes or
75 algorithms used to select the population should be referenced.”[10] Regarding AE-IPF,

one validation study has been performed in the US;[11] however, the positive predictive value (PPV) was not very high (0.621 [95% confidence interval [CI] 0.533 to 0.704]). Additionally, it remains unclear whether the data can be applied in other countries.

This study aimed to develop criteria for identifying patients with AE-IPF, as well as validate the criteria in the previous study, using multi-center chart data in Japan.

Methods

We performed and reported this study in accordance with the reporting guidelines for assessing the quality of validation studies of health administrative data (Table S1).[12]

Settings and patients

This retrospective, multicenter, validation study was conducted to verify the diagnostic accuracy of the disease name for AE-IPF. We performed study in eight Japanese tertiary hospitals: Saiseikai Kumamoto Hospital, Hyogo Prefectural Amagasaki General Medical Center, Iizuka Hospital, Kobe City Medical Center General Hospital, Kameda Medical Center, Hoshigaoka Medical Center, Okinawa Chubu Hospital, and the Japanese Red Cross Medical Center.

92 We included patients aged ≥ 40 years with a disease name that could meet the
93 diagnostic criteria for AE-IPF (Table 1), who were admitted to the aforementioned eight
94 institutes from January 2016 to February 2019. These criteria were developed in
95 advance by two respiratory specialists. We excluded patients with disease names for
96 secondary interstitial pneumonia, including collagen vascular disease-associated
97 interstitial lung disease and chronic and hypersensitivity pneumonitis, as well as those
98 for malignant neoplasms (Table 2). We did not exclude patients with disease names for
99 malignant neoplasms with the suffixes “post-operation” or “post-radiotherapy”. We
100 ignored all "suspected" disease names.

101 Regarding disease names, we extracted data from the Diagnosis Procedure Combination
102 (DPC), which was launched at Japanese acute-care hospitals across the country in
103 2003.[13, 14] The following data were extracted from the DPC: some clinical
104 information (e.g. age, gender, etc.), admission route (e.g. scheduled or emergency
105 admission), diagnoses, surgeries and procedures, medications, outcomes, etc. The
106 patients were recorded with diagnostic names using the International Statistical
107 Classification of Diseases and Related Health Problems, Tenth Revision (ICD-10)
108 codes. The diagnoses included the following six categories: main diagnosis, diagnosis
109 that triggered hospitalization, diagnosis using the most medical resources, diagnosis

using the second most medical resources, comorbidities present at admission, and conditions occurring after admission. One diagnostic name is coded for each main diagnosis, diagnosis that triggered hospitalization, diagnosis using the most medical resources, and diagnosis using the second most medical resources. Meanwhile, up to 10 diagnostic names can be coded for comorbidities present at admission and conditions occurring after admission. Furthermore, in the Japanese DPC data, modifiers can be added to the disease name in ICD-10 (e.g., regions such as left and right, recurrence, acute exacerbation, suspected, etc.).

This study was approved by the Ethics Committee of leading institute of this study (Kyoto University Graduate School and Faculty of Medicine) (Approved Number: R2071) and other committed institutes. The requirement for written informed consent from the participants was waived because of the retrospective design. Meanwhile, we offered the patients an opportunity to opt-out.

Reference standard

We confirmed the AE-IPF diagnosis using the following two steps:

- 1) At least two respiratory physicians at each institute reviewed the medical records and chest computed tomography (CT) findings on admission. Subsequently, they

127 determined whether the patients met the following diagnostic criteria: an official
128 ATS/ERS/JRS/ALAT clinical practice guideline for IPF diagnosis [15] and the
129 international working group diagnostic criteria for AE-IPF.[16] An IPF diagnosis was
130 determined if UIP or probable UIP pattern was observed on HRCT findings, even
131 without surgical lung biopsy (SLB). If SLB was performed, IPF was considered based
132 on specific combinations of HRCT and histopathological patterns following the
133 ATS/ERS/JRS/ALAT clinical practice guidelines.[15] Discrepancies were resolved
134 through discussion or consultation with a third respiratory physician, as appropriate.

135 2) Two expert chest radiologists interpreted chest CT findings of cases considered to be
136 AE-IPF in 1) and determined whether the patients met the aforementioned two
137 diagnostic criteria. If both radiologists considered that a case was not AE-IPF, it was
138 determined as not AE-IPF. On the other hand, if a case was considered as not as AE-IPF
139 by only one radiologist, the case was determined as AE-IPF.

140 Multidisciplinary discussion (MDD), which is held for diagnostic decision-making
141 regarding patients with interstitial lung disease who are clinically suspected to have IPF,
142 has been described in the official ATS/ERS/JRS/ALAT clinical practice guideline.[15]

However, MDD is a conditional recommendation that is not widely held in Japanese hospitals. Therefore, we decided not to consider MDD as a diagnostic requirement.

Developing criteria

First, we assessed the diagnostic accuracy of each disease name in the inclusion criteria, with disease names showing high diagnostic accuracy being adopted without modification. Next, we explored combinations of disease names with high diagnostic accuracy for disease names with a high number of true positives. We sought to create two criteria: 1) the criteria with the higher PPV (narrow criteria) and 2) criteria with slightly less PPV but more true positives (broad criteria). Based on previous studies, [11, 17, 18] we excluded disease names of secondary interstitial lung diseases (ILDs) and included those of acute respiratory failure (ARF) (Table 3). Further, we excluded disease names with pneumonia, influenza, and aspiration pneumonia and addition of that of IPF.

External validation of the criteria of a previous study

To compare the PPV with the aforementioned newly developed criteria, we externally validated the criteria of a previous study in the US [11] (pre-existing criteria) using DPC data extracted from the aforementioned eight hospitals. The inclusion criteria were

as follows: 1) age $\geq$ 50 years; 2) having at least one claim to a specific diagnostic code for IPF (either ICD10 code J84.9, J84.10, J84.111, J84.112, or ICD-9 correspond); 3) having acute respiratory symptoms or ARF (J84.114, J84.9, J96.00-02, J96.20-22, J96.91-92, or ICD-9 correspond) and subsequent hospital admission; and 4) lacking secondary ILDs.[11] In accordance with the Japanese DPC data used in this study, we modified this criteria as follows: 1) age $\geq$ 50 years; 2) having at least one following disease name (either ICD10 code J84.9 [interstitial pneumonia (IP)] or J84.1 [IPF or idiopathic interstitial pneumonia (IIP) or pulmonary fibrosis] in three diagnosis categories (main diagnosis, diagnosis that triggered hospitalization, diagnosis using the most medical resources); 3) having at least one following disease names (ICD10 code J84.1 [acute interstitial pneumonia] or J96.0 [ARF]) in any diagnosis category, and emergency admission; and 4) lacking secondary ILDs.

Statistical analysis

Interobserver agreement for the AE-IPF diagnosis between both respiratory specialists was evaluated using kappa statistics. For cases diagnosed as AE-IPF by two respiratory physicians, the interobserver agreement for AE-IPF diagnosis between both radiologists was assessed using the AC₁-index since most cases were considered to be true positive

cases.[19] We calculated the PPV as true positives / (true positives + false positives) for every disease name and their combinations. Further, we calculated the 95% confidence intervals (CIs) for binomial distribution using the exact method. We could not calculate the sensitivity, specificity, and negative predictive values since we did not perform a chart review of patients without target ICD-10 codes. Validation studies for only obtaining a PPV have reported that a sample size of approximately 100 is sufficient[20], because with that number of samples, the 95% CI falls within the range of the point estimate ± 0.1 . We sought to identify patients from various disease names and search for algorithms that would improve PPV. Therefore, regarding the narrow criteria, we identified the final criteria with the largest PPV out of approximately 100 cases. Statistical analyses were performed using STATA/SE version.16.0 (Stata Corp., College Station, TX, USA).

Results

This study included 822 patients; among them, 212 were diagnosed with AE-IPF by chart review. There was substantial interobserver agreement for AE-IPF diagnosis ($\kappa = 0.80$ [0.75 to 0.85]). Among the patients who were considered to have AE-IPF by both respiratory physicians, none were considered not to have AE-IPF by both

194 radiologists; moreover, there was excellent interobserver agreement between both
195 radiologists (AC_1 -index = 0.99 [0.98 to 1]).

196 Table 4 and Table S2 show the PPV and 95% CI for each disease name. Although the
197 disease name of AE-IPF and acute exacerbation of IIP (AE-IIP) had the highest and
198 second-highest PPV, respectively, the number of patients was small. The PPV for all the
199 other disease names was low.

200 **Narrow criteria**

201 As a result of examining each of the disease names, we found that the following five
202 disease names (AE-IIP, acute exacerbation of interstitial pneumonia, AE-IP, IPF +
203 emergency admission, IIP + emergency admission, and IP + emergency admission) in
204 three categories (main diagnosis, diagnosis that triggered hospitalization, or diagnosis
205 using the most medical resources) contained many true positive patients. Therefore, we
206 explored algorithms that would result in higher PPV for these disease names. We
207 attempted to exclude disease names related to secondary ILDs (any of all categories)
208 and infection (in four categories; main diagnosis, diagnosis that triggered
209 hospitalization, diagnosis using the most medical resources, or comorbidities present at
210 admission), and add to disease names for ARF (any of all categories) and IPF (any of all

211 categories). Consequently, we observed an increased PPV for several combinations of
212 disease names.

213 Finally, we combined the groups of disease names with a higher PPV and yielded a
214 criteria (AE-IPF OR (AE-IIP - secondary ILDs - infection) OR (AE-IIP + IPF) OR (AE-
215 IP - secondary ILDs - infection + ARF) with a PPV of 0.72 (95% CI 0.62 to 0.81)
216 (Table 4).

217 **Broad criteria**

218 Using the five disease names, we searched for a criteria that would include more true-
219 positive patients even with a slightly inferior PPV. Finally, we created a criteria (AE-
220 IPF OR AE-IIP OR [AE-IP - secondary ILDs + ARF] OR [IPF + emergency admission
221 + [ARF OR AE-IP] OR [IIP + emergency admission + (ARF OR AE-IP)] with a PPV
222 of 0.61 (95% CI 0.53 to 0.68) (Table 4).

223 **Pre-existing criteria**

224 The validation results for the pre-existing criteria (IPF OR IIP OR pulmonary fibrosis
225 OR IP) + (AIP OR ARF) + emergency admissions - secondary ILDs) showed a PPV of
226 0.40 (95% CI 0.31 to 0.49) (Table 4).

227 **Discussion**

228 Using Japanese administrative data, we created criteria for identifying patients with AE-
229 IPF and validated the pre-existing criteria. It was difficult to accurately identify true
230 positive AE-IPF cases only based on a single disease name. Consequently, we excluded
231 or added various other disease names and emergency admissions, which improved the
232 AE-IPF diagnosis accuracy. The pre-existing criteria had a relatively low PPV. The
233 PPV for the final criteria were as follows: narrow criteria, 0.72 (0.62 to 0.81); broad
234 criteria, 0.61 (0.53 to 0.68); and pre-existing criteria, 0.40 (0.31 to 0.49).

235 The diagnostic accuracy of the narrow and broad criteria showed a fair PPV.
236 Currently, the only validation study on AE-IPF derived a criteria with a PPV of 0.621
237 (95% CI 0.533 to 0.704),[11] which was lower and comparable to the PPV of the
238 narrow and broad criteria, respectively. Most disease name combinations in the narrow
239 criteria used the suffix acute exacerbation, which is unique to the DPC data and may
240 have attributed to the higher PPV compared with that in the previous study. Meanwhile,
241 the validation results for the pre-existing criteria revealed a relatively low PPV (0.40
242 [95% CI 0.31 to 0.49]). This indicates that it may be better to use the narrow or broad
243 criteria rather than the pre-existing criteria in the Japanese DPC data.

Meanwhile, our findings are suggestive of the difficulty of AE-IPF diagnosis only based on administrative data. As shown in Table 4, there were only four true-positive patients with a disease name of AE-IPF. IPF is a rare disease; moreover, its diagnosis is heavily dependent on high-resolution CT (HRCT). Additionally, IPF diagnosis is sometimes difficult since it requires the exclusion of all other diseases that cause chronic fibrosis of the lungs. Furthermore, in case of acute respiratory worsening in fibrotic ILDs, [21] it can be challenging for clinicians to distinguish between AE and others. Therefore, it is very difficult to accurately diagnose AE-IPF; moreover, many true positive patients could have been included among those with disease names of IIP or IP as well as IPF with or without AE. To exclude secondary ILDs, we referred to the ICD-9 and 10 codes used in previous IPF database studies.[11, 17, 18] For AE, we referred to a previous study and added a code for ARF.[11] Combining these codes with IIP and IP increased the PPV; therefore, these codes may be useful for excluding secondary ILDs and including AEs. For diseases that are rare and difficult to diagnose, a PPV of approximately $\geq 70\%$ is considered reasonable [22, 23]. Therefore, given the aforementioned difficulties in diagnosing AE-IPF, the PPVs obtained in our study could be considered reasonable.

Research implications

262 We believe our findings regarding the criteria for identifying patients with AE-IPF can
263 contribute to database research in Japan. Meanwhile, using the narrow criteria, only
264 about 1/3 of the cases were true positives. To detect more patients with AE-IPF, it may
265 be better to use the broad criteria even though it has a lower PPV. Additionally, the
266 narrow and broad criteria are both insufficient for selecting patients with AE-IPF
267 patients without omission. It should be recognized that some patients with disease
268 names, including acute respiratory distress syndrome or drug-induced interstitial
269 pneumonia, may meet the criteria for AE-IPF. Furthermore, the institutions involved in
270 this study were tertiary hospitals that treat numerous patients with AE-IPF. Although
271 DPC data are collected from numerous acute care hospitals, there were not all proficient
272 in treating patients with AE-IPF; thus, our findings may not be generalizable as
273 representative data for Japan.

274 We created criteria for defining AE-IPF with not very high PPV. Although the present
275 study and a previous validation study were conducted only using disease names and
276 hospitalization information, the DPC data have rich data on medications and
277 procedures. Therefore, by adding these information, such as information on
278 administration of systemic corticosteroid therapy, it may be possible to employ an
279 algorithm with a higher PPV. Additionally, using PPV alone cannot allow for a

database study that covers the entire AE-IPF population. Therefore, there is a need for studies calculating the sensitivity and specificity using methods, including random sampling or "all possible cases" sampling.[24]

Strength and Limitations

Our study has several strengths. First, this is the first validation study on AE-IPF using Japanese DPC data. Second, this was a multi-center study with a sufficient sample size. Third, AE-IPF diagnosis was more reliable because of the interpretation by two chest radiologists in addition to at least two pulmonologists.

This study had several limitations. First, since this study was conducted at tertiary emergency hospitals with respiratory specialists, it remains unclear whether it can be adapted to the entire DPC data in Japan. Therefore, our findings should be externally validated in other facilities in Japan. Second, this was a validation study of the Japanese DPC data, which is unique to Japan and does not apply to other countries. Finally, we cannot eliminate the possibility of misclassification of the IPF diagnosis (e.g., hypersensitivity pneumonitis and collagen vascular disease) or AE. However, we believe that this was reduced by the interpretation of the chest CT findings by two expert chest radiologists.

297 **Conclusion**

298 In conclusion, this study derived criteria for identifying AE-IPF from Japanese
299 administrative data with a fair PPV. When studying AE-IPF in Japanese administrative
300 data using the narrow or broad criteria, researchers should carefully interpret the target
301 population.

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320 **Author Contributions**

321 KA is the main author and contributed to the design of the project, data collection and
322 analysis, and wrote the draft of the manuscript. YK contributed to the design of the
323 project and data collection and analysis. KI, KK, TJ, KF, KT, RT, HI, TN, TK, and MI
324 contributed data collection. YY contributed to the design of the project and supervised
325 the study. All authors contributed to data interpretation, revised the draft, and reviewed
326 and approved the final manuscript.

327 **References**

328 1. du Bois RM. An earlier and more confident diagnosis of idiopathic pulmonary
329 fibrosis. *Eur Respir Rev* 2012;21:141–6.

- 330 2. Hutchinson J, Fogarty A, Hubbard R, McKeever T. Global incidence and
331 mortality of idiopathic pulmonary fibrosis: A systematic review. *Eur Respir J*
332 2015;46:795–806.
- 333 3. Hutchinson JP, McKeever TM, Fogarty AW, Navaratnam V, Hubbard RB.
334 Increasing global mortality from idiopathic pulmonary fibrosis in the twenty-first
335 century. *Ann Am Thorac Soc* 2014;11:1176–85.
- 336 4. Navaratnam V, Fogarty AW, Glendening R, McKeever T, Hubbard RB. The
337 increasing secondary care burden of idiopathic pulmonary fibrosis: hospital admission
338 trends in England from 1998 to 2010. *Chest* 2013;143:1078–84.
- 339 5. Richeldi L, du Bois RM, Raghu G, Azuma A, Brown KK, Costabel U, et al.
340 Efficacy and safety of nintedanib in idiopathic pulmonary fibrosis. *N Engl J Med*
341 2014;370:2071–82.
- 342 6. Song JW, Hong SB, Lim CM, Koh Y, Kim DS. Acute exacerbation of
343 idiopathic pulmonary fibrosis: incidence, risk factors and outcome. *Eur Respir J*
344 2011;37:356–63.

- 345 7. Raghu G, Collard HR, Egan JJ, Martinez FJ, Behr J, Brown KK, et al. An
346 official ATS/ERS/JRS/ALAT statement: Idiopathic pulmonary fibrosis: Evidence-based
347 guidelines for diagnosis and management. *Am J Respir Crit Care Med* 2011;183:788–
348 824.
- 349 8. Homma S, Bando M, Azuma A, Sakamoto S, Sugino K, Ishii Y, et al. Japanese
350 guideline for the treatment of idiopathic pulmonary fibrosis. *Respir Investig*
351 2018;56:268–91.
- 352 9. Schneeweiss S, Avorn J. A review of uses of health care utilization databases
353 for epidemiologic research on therapeutics. *J Clin Epidemiol* 2005;58:323–37.
- 354 10. Benchimol EI, Smeeth L, Guttman A, Harron K, Moher D, Petersen I, et al.
355 The REporting of studies Conducted using Observational Routinely-collected health
356 Data (RECORD) statement. *PLoS Med* 2015;12:e1001885.
- 357 11. Farrand E, Vittinghoff E, Ley B, Butte AJ, Collard HR. Corticosteroid use is
358 not associated with improved outcomes in acute exacerbation of IPF. *Respirology*
359 2020;25:629–35.

- 360 12. Benchimol EI, Manuel DG, To T, Griffiths AM, Rabeneck L, Guttman A.
361 Development and use of reporting guidelines for assessing the quality of validation
362 studies of health administrative data. *J Clin Epidemiol* 2011;64:821–9.
- 363 13. Okamura S, Kobayashi R, Sakamaki T. Case-mix payment in Japanese medical
364 care. *Health Policy* 2005;74:282–6.
- 365 14. Yasunaga H, Ide H, Imamura T, Ohe K. Impact of the Japanese Diagnosis
366 Procedure Combination-based Payment System on cardiovascular medicine-related
367 costs. *Int Heart J* 2005;46:855–66.
- 368 15. Raghu G, Jardin MR, Myers JL, Richeldi L, Ryerson CL, Lederer DJ, et al.
369 Diagnosis of idiopathic pulmonary fibrosis. An Official ATS/ERS/JRS/ALAT Clinical
370 Practice Guideline. *Am J Respir Crit Care Med* 2018;198:e44–e68.
- 371 16. Collard HR, Ryerson CJ, Corte TJ, Jenkins G, Kondoh Y, Lederer DJ et al.
372 Acute exacerbation of idiopathic pulmonary fibrosis. An international working group
373 report. *Am J Respir Crit Care Med* 2016;194:265–75.

- 374 17. Raghu G, Weycker D, Edelsberg J, Bradford WZ, Oster G. Incidence and
375 prevalence of idiopathic pulmonary fibrosis. *Am J Respir Crit Care Med* 2006;174:810–
376 6.
- 377 18. Lai CC, Wang CY, Lu HM, Chen L, Teng NC, Yan YH, et al., Idiopathic
378 pulmonary fibrosis in Taiwan - A population-based study. *Respir Med* 2012;106:1566–
379 74.
- 380 19. Shankar V, Bangdiwala SI. Observer agreement paradoxes in 2x2 tables:
381 Comparison of agreement measures. *BMC Med Res Methodol* 2014;14:100.
- 382 20. Cutrona SL, Toh S, Iyer A, Foy S, Cavagnaro E, Forrow S et al., Design for
383 validation of acute myocardial infarction cases in Mini-Sentinel. *Pharmacoepidemiol*
384 *Drug Saf* 2012;21:274–81.
- 385 21. Moua T, Westerly BD, Duloherly MM, Daniels CE, Ryu JH, Lim KG. Patients
386 with fibrotic interstitial lung disease hospitalized for acute respiratory worsening: A
387 large cohort analysis. *Chest* 2016;149:1205–14.

- 388 22. Segal JB, Powe NR. Accuracy of identification of patients with immune
389 thrombocytopenic purpura through administrative records: A data validation study. Am
390 J Hematol 2004;75:12–7.
- 391 23. Wahl PM, Terrell DR, George JN, Rodgers JK, Uhl L, Cataland S, et al.
392 Validation of claims-based diagnostic codes for idiopathic thrombotic
393 thrombocytopenic purpura in a commercially-insured population. Thromb Haemost
394 2010;103:1203–9.
- 395 24. Widdifield J, Ivers NM, Young J, Green D, Jaakkimainen L, Butt DA, et al.
396 Development and validation of an administrative data algorithm to estimate the disease
397 burden and epidemiology of multiple sclerosis in Ontario, Canada. Mult Scler
398 2015;21:1045–54.

Table 1

Disease names adopted as inclusion criteria

ICD-10 code 1	disease name 1*	suffix	ICD-10 code 2	disease name 2**	suffix	emergency admission
J841	idiopathic pulmonary fibrosis	AE	-	-	-	-
J841	idiopathic interstitial pneumonia	AE	-	-	-	-
J841	usual interstitial pneumonia	AE	-	-	-	-
J841	pulmonary fibrosis	AE	-	-	-	-
J841	diffuse interstitial pneumonia	AE	-	-	-	-
J849	interstitial pneumonia	AE	-	-	-	-
J841	acute interstitial pneumonia	-	-	-	-	-
J841	diffuse interstitial pneumonia	-	-	-	-	-
J841	diffuse alveolar damage	-	-	-	-	-
J80	acute respiratory distress syndrome	-	-	-	-	-
J984	acute lung injury	-	-	-	-	-
J702	acute drug-induced interstitial lung disorders	-	-	-	-	-
J704	drug-induced interstitial pneumonia	-	-	-	-	-
J10~J111 J15~189 J690	pneumonia, Influenza, aspiration pneumonia	-	J841	idiopathic pulmonary fibrosis	AE	-
		-	J841	idiopathic interstitial pneumonia	AE	-
		-	J841	usual interstitial pneumonia	AE	-
		-	J841	pulmonary fibrosis	AE	-
		-	J841	diffuse interstitial pneumonia	AE	-

		-	J841	interstitial pneumonia	AE	-
		-	J841	idiopathic pulmonary fibrosis	-	-
		-	J841	idiopathic interstitial pneumonia	-	-
		-	J841	usual interstitial pneumonia	-	-
		-	J841	pulmonary fibrosis	-	-
		-	J841	diffuse interstitial pneumonia	-	-
		-	J841	interstitial pneumonia	-	-
J841	idiopathic pulmonary fibrosis	-	-	-	-	○
J841	idiopathic interstitial pneumonia	-	-	-	-	○
J841	usual interstitial pneumonia	-	-	-	-	○
J841	pulmonary fibrosis	-	-	-	-	○
J841	diffuse interstitial pneumonia	-	-	-	-	○
J849	interstitial pneumonia	-	-	-	-	○

AE, acute exacerbation; DPC, diagnosis procedure combination; ICD-10, the International Statistical Classification of Diseases and Related Health Problems, Tenth Revision

*Disease name in any categories in the DPC data

**Disease name in co-morbidities present at admission and conditions occurring after admission

Table 2

Disease names adopted as exclusion criteria

ICD-10 code	disease name*
C00~C96	malignant neoplasms**
D860	sarcoidosis of lung
J60	coal worker's pneumoconiosis
J61	asbestosis
J64	pneumoconiosis
J67	hypersensitivity pneumonitis due to organic dust
J990 • M0510	rheumatoid arthritis-associated interstitial lung disease
J991 • M351	collagen vascular disease-associated interstitial lung disease
J991 • M330	juvenile dermatomyositis-associated interstitial lung disease
J991 • M321	systemic lupus erythematosus-associated interstitial lung disease
J991 • M332	polymyositis-associated interstitial lung disease
J991 • M331	dermatomyositis-associated interstitial lung disease
J991 • M313	lung involvement in granulomatosis with polyangiitis
M348	lung involvement in systemic sclerosis

* Disease name in any category in the DPC data

**not exclude if they have attached suffix of “post-operation” or “post-radiotherapy”.

Table 3

ICD-10 codes for interstitial lung diseases (ILDs) other than idiopathic pulmonary fibrosis (IPF) and acute respiratory failure.

ILDs other than IPF	
ICD-10 code	disease name
C966	Langerhans-cell histiocytosis*
D869	Sarcoidosis
E756	Lipidoses
J998 • E831	Idiopathic pulmonary hemosiderosis
E85	Amyloidosis
J60	coal worker's pneumoconiosis*
J61	asbestosis*
J62	silicosis or talcosis
J63	berylliosis and other inorganic dusts
J64	unspecified pneumoconiosis*
J67	hypersensitivity pneumonitis*
J82	Pulmonary eosinophilia, not elsewhere classified
J82	Extrinsic allergic alveolitis
J684	Chronic respiratory conditions due to chemicals, gases, fumes, and vapors
J701	Chronic and other pulmonary manifestations due to radiation
J708	Respiratory conditions due to other specified external agents
J840	Alveolar and parieto-alveolar conditions
J841	Postinflammatory pulmonary fibrosis
J99	Respiratory disorders in diseases classified elsewhere
K50	Regional enteritis
M05, M06	Rheumatoid arthritis
M310	Goodpasture' s syndrome
M313	Wegener' s granulomatosis
M317	Microscopic polyangiitis
M318	Other specified necrotizing vasculopathies
M321, M329	Systemic lupus erythematosus
M330, M331, M339	Dermatomyositis
M332	Polymyositis
M340, M341, M349	Systemic sclerosis
M348	Lung involvement in systemic sclerosis*

M350	Sjogren' s disease
M351	other overlap syndrome
M45	Ankylosing spondylitis
Q850	Neurofibromatosis
Q851	Tuberous sclerosis
Acute respiratory failure	
ICD-10 code	disease name
J960	acute respiratory failure

*We already excluded these disease names as exclusion criteria

Table 4

The positive predictive value (PPV) and 95% confidence intervals (CI) of patients diagnosed with acute exacerbation of idiopathic pulmonary fibrosis within each algorithm

disease names	total (n)	True Positives	PPV	95% CI
IPF + AE	5	4	0.8	0.28 to 0.99
IIP + AE	32	24	0.75	0.57 to 0.89
(IIP + AE) - secondary ILDs*	30	23	0.77	0.58 to 0.90
(IIP + AE) - secondary ILDs + ARF*	22	17	0.77	0.55 to 0.92
(IIP + AE) - secondary ILDs - infection	29	22	0.76	0.55 to 0.92
(IIP + AE) - secondary ILDs - infection + ARF	21	16	0.76	0.56 to 0.90
(IIP + AE) + IPF	6	6	1	0.54 to 1
IP + AE	105	59	0.56	0.46 to 0.66
(IP + AE) - secondary ILDs	95	57	0.6	0.49 to 0.70
(IP + AE) - secondary ILDs + ARF	75	50	0.67	0.55 to 0.77
(IP + AE) - secondary ILDs - infection	77	48	0.62	0.51 to 0.73
(IP + AE) - secondary ILDs - infection + ARF	60	42	0.7	0.57 to 0.81
(IP + AE) + IPF	15	9	0.6	0.32 to 0.84
(IP + AE) + IPF - secondary ILDs - infection	12	8	0.67	0.35 to 0.90
IPF + emergency admission	40	16	0.4	0.25 to 0.57
IPF + emergency admission + ARF	15	10	0.67	0.38 to 0.88
IPF + emergency admission - secondary ILDs - infection + ARF	12	8	0.67	0.35 to 0.90
IPF + emergency admission + (IP + AE)	4	2	0.5	0.068 to 0.93
IIP + emergency admission	152	35	0.23	0.17 to 0.31
IIP + emergency admission + ARF	47	19	0.40	0.26 to 0.56

IIP + emergency admission - secondary ILDs - infection + ARF	21	7	0.33	0.15 to 0.57
IIP + emergency admission + (IP + AE)	3	2	0.67	0.094 to 0.99
IP + emergency admission	140	38	0.27	0.20 to 0.35
IP + emergency admission + ARF	78	27	0.35	0.24 to 0.46
IP + emergency admission - secondary ILDs - infection + ARF	45	14	0.31	0.18 to 0.47
final algorithm				
Narrow criteria				
(IPF + AE) OR ((IIP + AE) - secondary ILDs - infection) OR ((IIP + AE) + IPF) OR ((IP + AE) - secondary ILDs - infection + ARF)	94	68	0.72	0.62 to 0.81
Broad criteria				
(IPF + AE) OR (IIP + AE) OR ((IP + AE) - secondary ILDs + ARF) OR (IPF + emergency admission + (ARF OR (IP + AE))) OR (IIP + emergency admission + (ARF OR (IP + AE)))	168	102	0.61	0.53 to 0.68
Pre-existing criteria				
((IPF OR IIP OR pulmonary fibrosis OR IP) + (AIP OR ARF) + emergency admission - secondary ILDs)	126	50	0.40	0.31 to 0.49

AE, acute exacerbation; AIP, acute interstitial pneumonia; ARF, acute respiratory failure; IIP, idiopathic interstitial pneumonia; ILDs, interstitial lung diseases; IP, interstitial pneumonia; IPF, idiopathic pulmonary fibrosis

“+” represent AND; “-“ represent NOT

*disease names of secondary ILDs and ARF are described in Table 3

Table S1. Checklist of reporting criteria for studies validating health administrative data algorithms.

	YES	NO	UNCERTAIN	NOT APPLICABLE
TITLE, KEYWORDS, ABSTRACT				
Identify article as study of assessing diagnostic accuracy	✓			
Identify article as study of administrative data	✓			
INTRODUCTION:				
State disease identification & validation one of goals of study	✓			
METHODS:				
<i>Participants in validation cohort:</i>				
Describe validation cohort (Cohort of patients to which reference standard was applied)	✓			
· Age		✓		
· Disease	✓			
· Severity				✓
· Location/Jurisdiction	✓			
Describe recruitment procedure of validation cohort	✓			
· Inclusion criteria	✓			
· Exclusion criteria	✓			

Describe patient sampling (random, consecutive, all, etc.)	✓			
Describe data collection	✓			
· Who identified patients and did selection adhere to patient recruitment criteria	✓			
· Who collected data	✓			
· <i>A priori</i> data collection form	✓			
· Disease classification				✓
· Split sample (i.e. re-validation using a separate cohort)				✓

	YES	NO	UNCERTAIN	NOT APPLICABLE
METHODS (cont.):				
<i>Test Methods:</i>				
Describe number, training and expertise of persons reading reference standard	✓			
If >1 person reading reference standard, quote measure of consistency (e.g. kappa)	✓			
Blinding of interpreters of reference standard to results of classification by administrative data e.g. Chart abstractor blinded to how that chart was coded			✓	
<i>Statistical Methods:</i>				

Describe methods of calculating/comparing diagnostic accuracy	✓			
RESULTS:				
<i>Participants:</i>				
Report when study done, start/end dates of enrollment	✓			
Describe number of people who satisfied inclusion/exclusion criteria	✓			
Study flow diagram				✓
<i>Test results:</i>				
Report distribution of disease severity				✓
Report cross-tabulation of index tests by results of reference standard	✓			

	YES	NO	UNCERTAIN	NOT APPLICABLE
RESULTS (cont.):				
<i>Estimates:</i>				
Report at least 4 estimates of diagnostic accuracy				✓
Diagnostic Accuracy Measures Reported:	✓			
· Sensitivity				✓
· Spec				✓
· PPV	✓			

· NPV				✓
· Likelihood ratios				✓
· Kappa	✓			
· Area under the ROC curve / c-statistic				✓
· Accuracy/agreement	✓			
· Other (specify)				✓
Report accuracy for subgroups (e.g. age, geography, different sex, etc.)				✓
If PPV/NPV reported, ratio of cases/controls of validation cohort approximate prevalence of condition in the population			✓	
Report 95% confidence intervals for each diagnostic measure	✓			
DISCUSSION:				
Discuss the applicability of the validation findings	✓			

Table S2

The positive predictive value (PPV) and 95% confidence intervals (CI) of patients diagnosed with acute exacerbation of idiopathic pulmonary fibrosis within each algorithm

disease names	total (n)	True Positives	PPV	95% CI
UIP + AE	0	0	-	-
pulmonary fibrosis + AE	2	1	0.5	0.13 to 0.99
diffuse IP + AE	8	1	0.13	0.003 to 0.53
AIP	40	11	0.28	0.15 to 0.44
diffuse IP	86	8	0.093	0.041 to 0.18
DAD	1	0	0	0 to 0.98
ARDS	66	5	0.076	0.025 to 0.17
ALI	2	0	0	0 to 0.84
acute drug-induced interstitial lung disorders	13	1	0.077	0.002 to 0.36
drug-induced interstitial pneumonia	47	2	0.043	0.005 to 0.15
Infection + (IPF + AE)	0	0	-	-
Infection + (IIP + AE)	1	0	0	0 to 0.98
Infection + (UIP + AE)	0	0	-	-
Infection + (pulmonary fibrosis + AE)	0	0	-	-
Infection + (diffuse IP+AE)	0	0	-	-
Infection + (IP + AE)	11	4	0.36	0.11 to 0.69
Infection + IPF	15	3	0.2	0.043 to 0.48
Infection + IIP	15	3	0.2	0.043 to 0.48
Infection + UIP	0	0	-	-
Infection + pulmonary fibrosis	5	0	0	0 to 0.52

Infection + diffuse IP	3	0	0	0 to 0.71
Infection + IP	82	11	0.13	0.069 to 0.23
UIP + emergency admission	11	4	0.36	0.11 to 0.69
pulmonary fibrosis + emergency admission	3	0	0	0 to 0.71
diffuse IP + emergency admission	53	7	0.13	0.055 to 0.25

AE, acute exacerbation; AIP, acute interstitial pneumonia; ALI, acute lung injury; ARDS, acute respiratory distress syndrome; DAD, diffuse alveolar damage; IIP, idiopathic interstitial pneumonia; IP, interstitial pneumonia; IPF, idiopathic pulmonary fibrosis; UIP, usual interstitial pneumonia

“+” represent AND; “-“ represent NOT

Table S3

Japanese version of AE-IPF algorithm using diagnosis procedure combination data

Specificity maximizing algorithm
<p>1) 入院時の年齢が 40 歳以上</p> <p>2) 以下の①～④のいずれかの病名が当てはまる。ただし、⑤の病名がある場合、あらかじめ除外しておく。</p> <p>①入院病名の主病名または入院契機病名または医療資源を最も投入した傷病に、以下のいずれかの病名がある。</p> <p>J841 (ICD-10) 「特発性肺線維症」+接頭語「急性増悪」</p> <p>②1～2 の病名の組み合わせを満たす</p> <p>1. <u>入院病名の主病名または入院契機病名または医療資源を最も投入した傷病に、以下の病名がある。</u></p> <p>J841 (ICD-10) 「特発性間質性肺炎」+接頭語「急性増悪」</p> <p>2. <u>入院病名のいずれかに以下の病名がある。</u></p> <p>J841 (ICD-10) 「特発性肺線維症」</p> <p>③1 の病名を満たし、2～3 に当てはまる病名を除外する</p> <p>1. <u>入院病名の主病名または入院契機病名または医療資源を最も投入した傷病に、以下の病名がある。</u></p> <p>J841 (ICD-10) 「特発性間質性肺炎」+接頭語「急性増悪」</p> <p>2. <u>入院病名の主病名または入院契機病名または医療資源を最も投入した傷病または入院時併存症に、以下のいずれかの病名がある。</u></p> <p>J101～J111 (ICD-10) 「インフルエンザ」</p> <p>J150～J189 (ICD-10) に該当する肺炎</p> <p>J690 (ICD-10) 「誤嚥性肺炎」</p> <p>3. <u>入院病名のいずれかに以下のいずれかの病名がある。</u></p> <p>C966 (ICD-10) 「ランゲルハンス細胞組織球症」*</p> <p>D869 (ICD-10) 「サルコイドーシス」</p> <p>E756 (ICD-10) 「リピドーシス」</p> <p>E831・J998 (ICD-10) 「特発性肺へモジデローシス」</p> <p>E85 (ICD-10) 「アミロイドーシス」</p>

J60 (ICD-10) 「炭坑夫塵肺症」＊
J61 (ICD-10) 「石綿肺」＊
J62 (ICD-10) 「珪酸を含む粉塵による塵肺症」
J63 (ICD-10) 「その他の無機粉塵による塵肺症」
J64 (ICD-10) 「塵肺症」＊
J67 (ICD-10) 「有機粉塵による過敏性肺臓炎」＊
J82 (ICD-10) 「肺好酸球症, 他に分類されないもの」
J684 (ICD-10) 「化学物質, ガス, フェーム及び蒸気による慢性呼吸器病態」
J701 (ICD-10) 「放射線による慢性及びその他の肺症状」
J708 (ICD-10) 「その他の明示された外的因子による呼吸器病態」
J840 (ICD-10) 「肺胞性及び肺胞周囲性病態」
J841 (ICD-10) 「炎症後肺線維症」
J99 (ICD-10) 「他に分類される疾患における呼吸器障害」
K50 (ICD-10) 「クローン病 [限局性腸炎]」
M05 (ICD-10) 「血清反応陽性関節リウマチ」
M06 (ICD-10) 「その他の関節リウマチ」
M310 (ICD-10) 「グッドパスチャー症候群」
M313 (ICD-10) 「Wegener 肉芽腫」
M317 (ICD-10) 「顕微鏡的多発血管炎」
M318 (ICD-10) 「その他の明示された壊死性血管障害」
M321, M329 (ICD-10) 「全身性エリテマトーデス」
M330, M331, M339 (ICD-10) 「皮膚筋炎」
M332 (ICD-10) 「多発性筋炎」
M340, M341, M349 (ICD-10) 「全身性硬化症」
M348 (ICD-10) 「全身性強皮症性呼吸器障害」＊

M350 (ICD-10) 「シェーグレン症候群」

M351 (ICD-10) 「その他の重複症候群」

M45 (ICD-10) 「強直性脊椎炎」

Q850 (ICD-10) 「神経線維腫症」

Q851 (ICD-10) 「結節性硬化症」

④1～2の病名の組み合わせを満たし、3～4に当てはまる病名を除外する

1. 入院病名の主病名または入院契機病名または医療資源を最も投入した傷病に、以下の病名がある。

J849 (ICD-10) 「間質性肺炎」+接頭語「急性増悪」

2. 入院病名のいずれかに以下の病名がある。

J960 (ICD-10) 「急性呼吸不全」

3. 入院病名の主病名または入院契機病名または医療資源を最も投入した傷病または入院時併存症に、以下のいずれかの病名がある。

J101～J111 (ICD-10) 「インフルエンザ」

J150～J189 (ICD-10) に該当する肺炎

J690 (ICD-10) 「誤嚥性肺炎」

4. 入院病名のいずれかに以下のいずれかの病名がある。

C966 (ICD-10) 「ランゲルハンス細胞組織球症」*

D869 (ICD-10) 「サルコイドーシス」

E756 (ICD-10) 「リピドーシス」

E831・J998 (ICD-10) 「特発性肺へモジデローシス」

E85 (ICD-10) 「アミロイドーシス」

J60 (ICD-10) 「炭坑夫塵肺症」*

J61 (ICD-10) 「石綿肺」*

J62 (ICD-10) 「珪酸を含む粉塵による塵肺症」

J63 (ICD-10) 「その他の無機粉塵による塵肺症」

J64 (ICD-10) 「塵肺症」*

J67 (ICD-10) 「有機粉塵による過敏性肺臓炎」＊
J82 (ICD-10) 「肺好酸球症, 他に分類されないもの」
J684 (ICD-10) 「化学物質, ガス, フューム及び蒸気による慢性呼吸器病態」
J701 (ICD-10) 「放射線による慢性及びその他の肺症状」
J708 (ICD-10) 「その他の明示された外的因子による呼吸器病態」
J840 (ICD-10) 「肺胞性及び肺胞周囲性病態」
J841 (ICD-10) 「炎症後肺線維症」
J99 (ICD-10) 「他に分類される疾患における呼吸器障害」
K50 (ICD-10) 「クローン病 [限局性腸炎]」
M05 (ICD-10) 「血清反応陽性関節リウマチ」
M06 (ICD-10) 「その他の関節リウマチ」
M310 (ICD-10) 「グッドパスチャー症候群」
M313 (ICD-10) 「Wegener 肉芽腫」
M317 (ICD-10) 「顕微鏡的多発血管炎」
M318 (ICD-10) 「その他の明示された壊死性血管障害」
M321, M329 (ICD-10) 「全身性エリテマトーデス」
M330, M331, M339 (ICD-10) 「皮膚筋炎」
M332 (ICD-10) 「多発性筋炎」
M340, M341, M349 (ICD-10) 「全身性硬化症」
M348 (ICD-10) 「全身性強皮症性呼吸器障害」＊
M350 (ICD-10) 「シェーグレン症候群」
M351 (ICD-10) 「その他の重複症候群」
M45 (ICD-10) 「強直性脊椎炎」
Q850 (ICD-10) 「神経線維腫症」
Q851 (ICD-10) 「結節性硬化症」

⑤入院病名の主傷病または入院契機病名または医療資源を最も投入した傷病または入院時併存症に以下の病名がある(あらかじめ除外する)。
ただし、癌（C000～C759・C800・C809・C81～C97）については、術後、放射線治療後の病名は除外しない。

C000～C759（ICD-10）「癌」

C800・C809（ICD-10）「原発不明癌」

C81～C97（ICD-10）「リンパ組織、造血組織及び関連組織の癌」

D860（ICD-10）「肺サルコイドーシス」

M0510・J990（ICD-10）「関節リウマチ性間質性肺炎」

M313・J991（ICD-10）「多発血管炎性肉芽腫症性呼吸器障害」

M321・J991（ICD-10）「全身性エリテマトーデス性間質性肺炎」

M330・J991（ICD-10）「若年性皮膚筋炎性間質性肺炎」

M331・J991（ICD-10）「皮膚筋炎性間質性肺炎」

M332・J991（ICD-10）「多発性筋炎性間質性肺炎」

M348（ICD-10）「全身性強皮症性呼吸器障害」

M351・J991（ICD-10）「膠原病性間質性肺炎」

J60（ICD-10）「炭坑夫塵肺症」

J61（ICD-10）「石綿肺」

J64（ICD-10）「塵肺症」

J67（ICD-10）「有機粉塵による過敏性肺臓炎」

Balanced algorithm

1) 入院時の年齢が 40 歳以上

2) 以下の①～③のいずれかの病名が当てはまる。ただし、④の病名がある場合、あらかじめ除外しておく。

①入院病名の主病名または入院契機病名または医療資源を最も投入した傷病に、以下のいずれかの病名がある。

J841（ICD-10）「特発性肺線維症」+接頭語「急性増悪」

J841（ICD-10）「特発性間質性肺炎」+接頭語「急性増悪」

②1～2 の病名の組み合わせを満たし、3 に当てはまる病名を除外する

1. 入院病名の主病名または入院契機病名または医療資源を最も投入した傷病に、以下の病名がある。

J849 (ICD-10) 「間質性肺炎」+接頭語「急性増悪」

2. 入院病名のいずれかに以下の病名がある。

J960 (ICD-10) 「急性呼吸不全」

3. 入院病名のいずれかに以下のいずれかの病名がある。

C966 (ICD-10) 「ランゲルハンス細胞組織球症」*

D869 (ICD-10) 「サルコイドーシス」

E756 (ICD-10) 「リピドーシス」

E831・J998 (ICD-10) 「特発性肺ヘモジデローシス」

E85 (ICD-10) 「アミロイドーシス」

J60 (ICD-10) 「炭坑夫塵肺症」*

J61 (ICD-10) 「石綿肺」*

J62 (ICD-10) 「珪酸を含む粉塵による塵肺症」

J63 (ICD-10) 「その他の無機粉塵による塵肺症」

J64 (ICD-10) 「塵肺症」*

J67 (ICD-10) 「有機粉塵による過敏性肺臓炎」*

J82 (ICD-10) 「肺好酸球症, 他に分類されないもの」

J684 (ICD-10) 「化学物質, ガス, フェューム及び蒸気による慢性呼吸器病態」

J701 (ICD-10) 「放射線による慢性及びその他の肺症状」

J708 (ICD-10) 「その他の明示された外的因子による呼吸器病態」

J840 (ICD-10) 「肺胞性及び肺胞周囲性病態」

J841 (ICD-10) 「炎症後肺線維症」

J99 (ICD-10) 「他に分類される疾患における呼吸器障害」

K50 (ICD-10) 「クローン病 [限局性腸炎]」

M05 (ICD-10) 「血清反応陽性関節リウマチ」

M06 (ICD-10) 「その他の関節リウマチ」
M310 (ICD-10) 「グッドパスチャー症候群」
M313 (ICD-10) 「Wegener 肉芽腫」
M317 (ICD-10) 「顕微鏡的多発血管炎」
M318 (ICD-10) 「その他の明示された壊死性血管障害」
M321, M329 (ICD-10) 「全身性エリテマトーデス」
M330, M331, M339 (ICD-10) 「皮膚筋炎」
M332 (ICD-10) 「多発性筋炎」
M340, M341, M349 (ICD-10) 「全身性硬化症」
M348 (ICD-10) 「全身性強皮症性呼吸器障害」*
M350 (ICD-10) 「シェーグレン症候群」
M351 (ICD-10) 「その他の重複症候群」
M45 (ICD-10) 「強直性脊椎炎」
Q850 (ICD-10) 「神経線維腫症」
Q851 (ICD-10) 「結節性硬化症」

③1～2 の組み合わせを満たす

1. 入院病名の主病名または入院契機病名または医療資源を最も投入した傷病に、以下のいずれかの病名があり、入院経路が緊急入院である。

J841 (ICD-10) 「特発性肺線維症」
J841 (ICD-10) 「特発性間質性肺炎」

2. 入院病名のいずれかに以下のいずれかの病名がある。

J960 (ICD-10) 「急性呼吸不全」
J849 (ICD-10) 「間質性肺炎」+接頭語「急性増悪」

④入院病名の主傷病または入院契機病名または医療資源を最も投入した傷病または入院時併存症に以下の病名がある(あらかじめ除外する)。
ただし、癌 (C000～C759・C800・C809・C81～C97) については、術後、放射線治療後の病名は除外しない。

C000～C759 (ICD-10) 「癌」
C800・C809 (ICD-10) 「原発不明癌」
C81～C97 (ICD-10) 「リンパ組織、造血組織及び関連組織の癌」
D860 (ICD-10) 「肺サルコイドーシス」
M0510・J990 (ICD-10) 「関節リウマチ性間質性肺炎」
M313・J991 (ICD-10) 「多発血管炎性肉芽腫症性呼吸器障害」
M321・J991 (ICD-10) 「全身性エリテマトーデス性間質性肺炎」
M330・J991 (ICD-10) 「若年性皮膚筋炎性間質性肺炎」
M331・J991 (ICD-10) 「皮膚筋炎性間質性肺炎」
M332・J991 (ICD-10) 「多発性筋炎性間質性肺炎」
M348 (ICD-10) 「全身性強皮症性呼吸器障害」
M351・J991 (ICD-10) 「膠原病性間質性肺炎」
J60 (ICD-10) 「炭坑夫塵肺症」
J61 (ICD-10) 「石綿肺」
J64 (ICD-10) 「塵肺症」
J67 (ICD-10) 「有機粉塵による過敏性肺臓炎」