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Incidence and Prevalence of Idiopathic Inflammatory Myopathies in Korea: a Nationwide Population-based Study

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ABSTRACT

Background: This study aimed to estimate the incidence and prevalence of idiopathic inflammatory myopathies (IIM) and associated comorbidities in Korea from 2006 to 2015.

Methods: IIM between 2004 to 2015 were identified using the Korean National Health Insurance Service medical claim database. The case definition required more than one visit based on diagnostic codes including juvenile dermatomyositis (JDM), dermatomyositis (DM), or polymyositis (PM) and registration in the Individual Copayment Beneficiaries Program (ICBP) for rare and intractable diseases. IIM patients with a disease-free period of 24 months before the index date were defined as incident cases. The Elixhauser comorbidity score was calculated.

Results: Using the base case definition, 1,150 prevalent patients with IIM (117 JDM, 521 DM, 512 PM) were recorded in 2006 and 2,210 (130 JDM, 1,101 DM, 869 PM) in 2015. The prevalence was estimated at 2.3–4.0 (0.9–1.2 for JDM, 1.2–2.7 for DM, 1.4–2.1 for PM)/100,000 person-year (PY). We identified 218 incident cases of IIM in 2006 (18 JDM, 98 DM, 102 PM) and 191 cases (7 JDM, 83 DM, 101 PM) in 2015. The incidence was estimated at 2.9–5.2 (0.7–1.9 for JDM, 1.8–4.0 for DM, 1.6–3.0 for PM)/1,000,000 PY. The mean age (\pm standard deviation) of prevalent patients with IIM was 51.2 (\pm 16.9) years, and the percentage of women was 72.1%. More than two-thirds of patients (70.7%) had more than two comorbidities. Twenty percent of patients had interstitial lung diseases.

Conclusion: In Korea, the incidence and prevalence of IIM were 2.9–5.2/1,000,000 PY and 2.3–4.0/100,000 PY, respectively.

Keywords: Inflammatory Myopathy; Prevalence; Incidence; Comorbidity

INTRODUCTION

Idiopathic inflammatory myopathies (IIM) have classical clinical manifestations of muscle weakness related to chronic inflammation in skeletal and systemic inflammation in other organs, including the skin, joints, lungs, gastrointestinal tract, and heart. On the basis of muscular symptoms, skin rash, and histopathological features, different subgroups have been identified with dermatomyositis (DM), polymyositis (PM), and inclusion body myositis.¹

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Disclosure

The authors have no potential conflicts of interest to disclose.

Author Contributions

Conceptualization: Cho SK, Jung SY, Yoo DH, Sung YK. Data curation: Myung J, Nam E, Jang EJ. Formal analysis: Cho SK, Kim H, Myung J, Nam E. Investigation: Kim H, Myung J, Jung SY, Jang EJ, Yoo DH, Sung YK. Methodology: Cho SK, Kim H, Myung J, Nam E, Jung SY, Jang EJ, Sung YK. Writing - original draft: Cho SK, Sung YK. Writing - review & editing: Kim H, Myung J, Nam E, Jung SY, Jang EJ, Yoo DH, Sung YK.

IIM are believed to be very rare, and their epidemiologic features have been poorly studied. A recent systematic literature review found that the incidence of IIM ranged from 1.16 to 19 incident cases/million/year and the prevalence from 2.4 to 33.8/100,000 persons.² The average onset age of IIM patients varies from 44.2 to 55.1 years, and women are dominant.²⁻⁴ The prevalence of DM increases significantly with geographical latitude from northern Europe to southern Europe,⁵ highlighting the potential role of environmental factors in disease development. The wide reported ranges in epidemiologic studies may be related to genetic and environmental factors, but the relationships are not well defined. In addition, there may also be differences in database, study design, heterogeneity in case ascertainment, and methodology. Therefore, an epidemiologic study of these rare diseases using a large population database is essential to identify geographical and population-based disparities and offer clues to disease characteristics and etiology.

Identification of the exact prevalence of IIM provides information on the disease burden based not only on number of patients, but also on comorbidities. The impact of comorbidities on disease outcome and prevalence has been well evaluated in other rheumatic diseases, especially rheumatoid arthritis (RA).^{6,7} IIM is considered to be associated with severe comorbidity, primarily related to muscle weakness and various systemic organ involvement, as well as increased mortality.⁸⁻¹¹ However, little is known about comorbidities related to IIM.

Therefore, we aimed to estimate the prevalence and incidence of IIM in Korea from 2006 to 2015 and to describe comorbidities in patients with IIM.

METHODS**Data source***Korean National Health Insurance Service (NHIS) database*

The entire Korean population in 2015 was 49,705,663 people. All people in Korea are eligible for coverage under the National Health Insurance Program. Therefore, a total number of 47 million, or over 96.3% of the total population, was included in the NHIS database.¹² The NHIS database contains individual beneficiary information, in addition to healthcare service information such as diagnosis, procedures, prescriptions, and tests. We used the NHIS database between January 2004 and December 2015.

Study population and covariates*Definition of IIM*

The case definition required more than one visit based on the myositis International Classification of Disease-tenth (ICD-10) diagnostic codes of M330, M331 or M339, M332, and registration with the national rare intractable disease supporting program. Patients who suffer from rare incurable diseases can be registered to the Individual Copayment Beneficiaries Program (ICBP) to reduce their burden of medical expenses.¹³ Application to this program for DM and PM requires a thorough clinical and laboratory survey that fulfills the diagnostic criteria proposed by Bohan and Peter.^{14,15} Patients' selection flow presented in the **Supplementary Fig. 1**.

Sub-diagnosis was classified as juvenile dermatomyositis (JDM, M330), DM (M331 or M339), and PM (M332). For patients with more than two different diagnostic codes of IIM at different

visits, sub-diagnosis was set by prioritizing the diagnostic code first registered to the ICBP. Adult patients aged more than 18 years with a JDM code were classified with the adult diagnostic code (DM or PM) from the closest visit date. However, adult patients with the JDM code applied continuously was classified with JDM. Juvenile patients aged less than 18 years were classified as JDM to avoid underestimation of juvenile myositis.

Prevalence and incidence

Prevalence cases were evaluated for each year between 2006 and 2015. To identify incident cases, patients without any diagnostic code or ICBP code for IIM for 24 months before the index date in each year were extracted. Patients older than 18 years with JDM were excluded for detection of incidence cases, because they may have previously been diagnosed with JDM but be in clinical remission.

Comorbidities

Comorbidities were detected based on the Elixhauser comorbidity index¹⁶ and calculated by ICD-10 codes within one year preceding the IIM index date in 2015 prevalent cases. In addition, pulmonary diseases such as interstitial lung disease, pneumothorax, pneumomediastinum, and acute myocardial infarction were also detected based on ICD-10 codes.

Statistical analysis

The overall prevalence per 100,000 persons and incidence per 1,000,000 persons between 2006 and 2015 were estimated. The year-specific mid-year population from the same year provided by the Korean National Statistics Office was used to calculate the prevalence and incidence of IIM. Categorical variables are presented as frequency and percentage, and continuous variables are presented as mean with standard deviation (SD) or median with interquartile ranges (IQR). All analyses were performed using SAS 9.2 (SAS Institute, Cary, NC, USA).

Ethics statement

This study was approved by the Institutional Review Board (IRB) of Hanyang University Medical Center and was exempt from an IRB review because we used existing, publicly available data and the information of the subjects could not be identified directly or through identifiers linked to the subjects (IRB No. HYUH 2017-09-009). Informed consent was waived because we utilized a de-identified database which was opened to the public.

RESULTS

Prevalence and incidence rate of IIM patients

Prevalence was estimated at 2.3–4.0 (0.9–1.2 for JDM, 1.4–2.7 for DM, 1.4–2.1 for PM)/100,000 persons (**Fig. 1**). We identified 1,150 prevalent cases of IIM in 2006 and 2100 cases in 2015. The mean age (\pm SD) of prevalent patients with IIM was 51.2 (\pm 16.9) years, and 72.1% were women. Most (82.4%) visited tertiary or general hospitals, with 74.5% of patients visiting internal medicine departments and 11.1% of patients visiting neurology departments. Among DM patients, 83.6% frequently visited internal medicine, 4.8% visited neurology, and 4.5% visited dermatology, while 70.9% of PM patients frequently visited internal medicine and 20.5% visited neurology. About 10% of patients with IIM visited an emergency room once per year, and 9.6% experienced hospitalizations three or more times per year (**Table 1**). Health care utilization was not different between DM and PM.

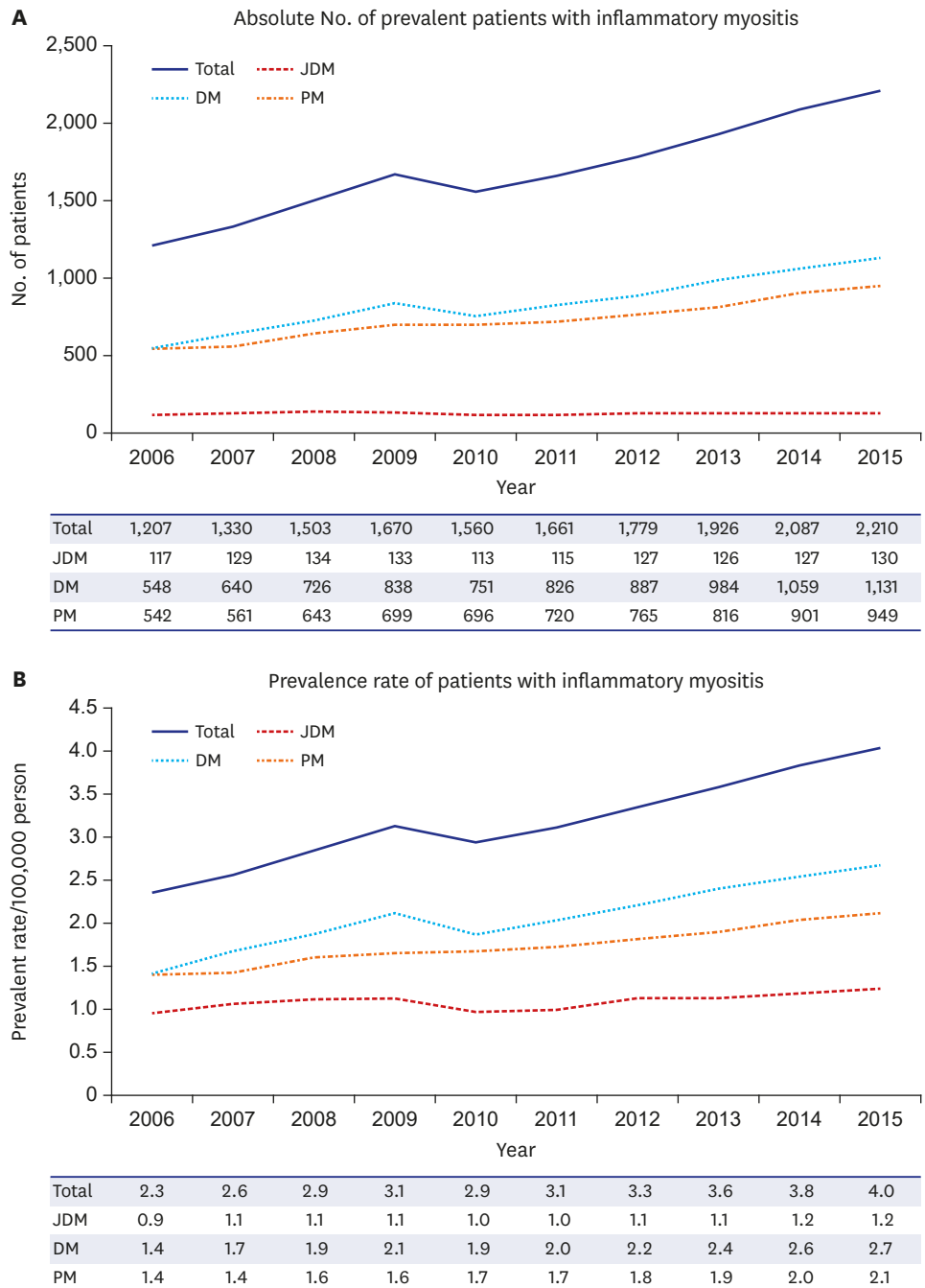


Fig. 1. Prevalence trends between 2006 and 2015 in patients with inflammatory myositis. JDM = juvenile dermatomyositis, PM = polymyositis, DM = dermatomyositis.

Table 1. Characteristics of prevalent patients with inflammatory myositis in 2015

Characteristics	Total (n = 2,100)	JDM (n = 130)	DM (n = 1,101)	PM (n = 869)
Age, yr (mean ± SD)	51.2 ± 16.9	12.7 ± 7.1	51.6 ± 14.1	56.5 ± 13.6
Gender, women	1,515 (72.1)	68 (52.3)	802 (72.8)	645 (74.2)
Insurance type				
National insurance	1,962 (93.4)	124 (95.4)	1,033 (93.8)	805 (92.6)
Medical aid	138 (6.6)	6 (4.6)	68 (6.2)	64 (7.4)
Type of institution ^a				
Tertiary hospital	1,550 (76.8)	99 (83.2)	838 (79.1)	613 (73.1)
General hospital	315 (15.6)	14 (11.8)	154 (14.5)	147 (17.5)
Hospital	49 (2.4)	2 (1.7)	23 (2.2)	24 (2.9)
Clinic	76 (3.8)	4 (3.4)	36 (3.4)	36 (4.3)
Others	28 (1.4)	-	9 (0.9)	19 (2.3)
Type of department ^a				
Internal medicine	1,504 (74.5)	23 (19.3)	886 (83.6)	595 (70.9)
Neurology	223 (11.1)	-	51 (4.8)	172 (20.5)
Dermatology	52 (2.6)	4 (3.4)	48 (4.5)	-
Pediatrics	90 (4.5)	77 (64.7)	12 (1.1)	1 (0.1)
Rehabilitation	44 (2.2)	7 (5.9)	14 (1.3)	23 (2.7)
Orthopedics	52 (2.6)	4 (3.4)	28 (2.6)	20 (2.4)
Others	53 (2.6)	4 (3.4)	21 (2.0)	28 (3.3)
The frequency of outpatient visit ^b (mean ± SD)	7.6 ± 3.9	7.6 ± 4.4	7.8 ± 3.9	7.4 ± 3.7
The frequency of admission ^b				
0	1,579 (75.2)	97 (74.6)	839 (76.2)	672 (77.3)
1	226 (10.8)	12 (9.2)	118 (10.7)	96 (11.1)
2	94 (4.5)	5 (3.9)	49 (4.5)	40 (4.6)
≥ 3	201 (9.6)	16 (12.3)	95 (8.6)	61 (7.0)
The frequency of ER visit ^b				
0	1,876 (89.3)	118 (90.8)	979 (88.9)	779 (89.6)
1	141 (6.7)	6 (4.6)	70 (6.4)	65 (7.5)
2	46 (2.2)	5 (3.9)	26 (2.4)	15 (1.7)
≥ 3	37 (1.8)	1 (0.8)	26 (2.4)	10 (1.2)

Data are presented as number (%).

JDM = juvenile dermatomyositis, DM = dermatomyositis, PM = polymyositis, SD = standard deviation, ER = emergency room.

^aFrequently visited over 1 year; ^bThe number of visits for 1 year.

We identified 218 incident cases of IIM in 2006 and 191 cases in 2015 (**Fig. 2A**). Incidence rates were estimated at 2.9–5.2 (0.7–1.9 for JDM, 1.8–4.0 for DM, 1.6–3.0 for PM)/1,000,000 person-year. The trend of IIM incidence rates did not appear to increase significantly over a period of 10 years (**Fig. 2B**).

Comorbidities of IIM patients

In 2015, the median with IQR of the Elixhauser comorbidity index was 3 (2, 5) in PM, and 2 (1, 4) in DM. More than two-thirds of patients (70.7%) had more than two comorbidities. Chronic pulmonary disease and uncomplicated hypertension were common among IIM patients, with prevalence rates of 34.1% and 27.1%, respectively, followed by peptic ulcer disease at 21.8%. In all sub-diagnosis, chronic pulmonary diseases were the most common comorbidity (**Fig. 3**). Uncomplicated hypertension, peptic ulcer disease, and liver diseases were followed in patients with DM, or PM. However, anemia was more common than liver disease and peptic ulcer disease in JDM.

The percentage of patients with ILD was 20.0% in IIM patients and was similar in DM and PM. However, pneumothorax (0.6% in DM vs. 0.4% in PM) and pneumomediastinum (0.8% in only DM) were more frequent in DM (**Table 2**). The percentage of patients with acute myocardial infarction was 1.9% in DM and 3.0% in PM.

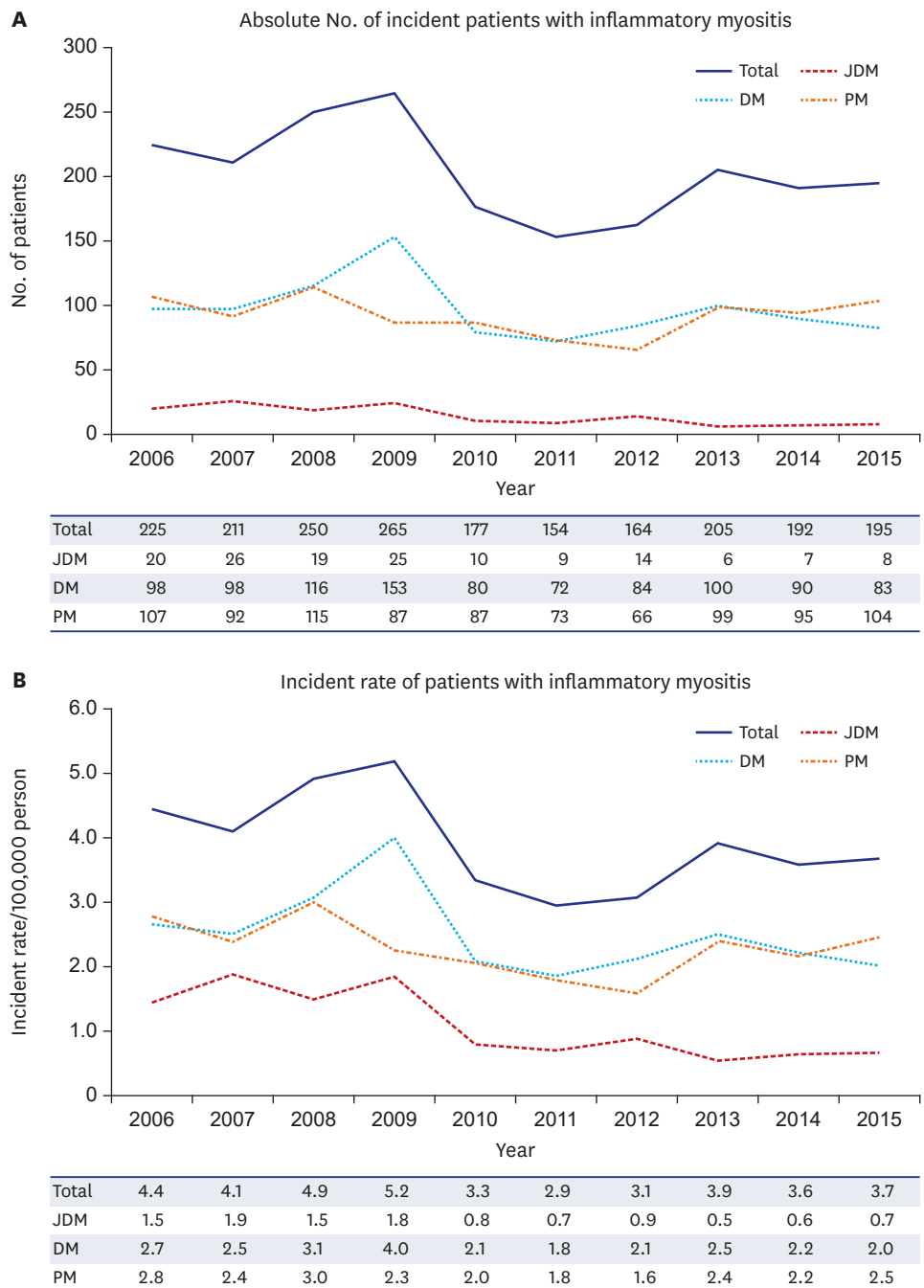


Fig. 2. Incidence trends between 2006 and 2015 in patients with inflammatory myositis. JDM = juvenile dermatomyositis, PM = polymyositis, DM = dermatomyositis.

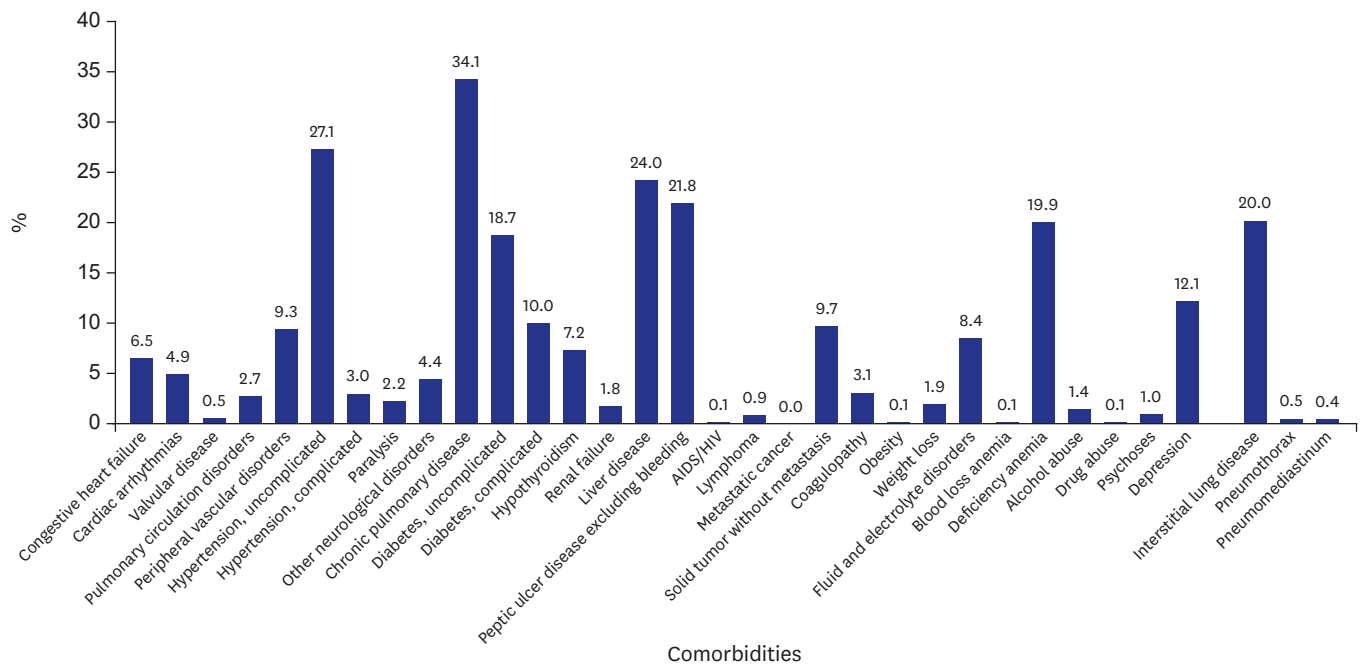


Fig. 3. Percentage of comorbidities of prevalent patients with inflammatory myositis in 2015. AIDS/HIV = acquired immune deficiency syndrome/human immunodeficiency virus.

DISCUSSION

Using a nationwide population-based database that included most of the Korean population, we estimated an overall IIM prevalence rate of 2.3–4.4/100,000 persons and an incidence of 2.9–5.2 per 1,000,000. We did not observe any trends in incidence over a 10-year period.

Our calculated prevalence and incidence were lower than those reported in a recent study from Sweden between 2007 and 2011 (prevalence of 14/100,000 and incidence of 11/1,000,000) and a meta-analysis of prevalence (14/100,000 between 1982 and 2010 and incidence 7.98/1,000,000 between 1951 and 2010). In comparison with the data from other Asian populations, our prevalence was similar to that of Taiwan at 2.9/100,000¹⁷ and lower than the reported Japanese prevalence of 13.2/100,000 using a registration system from an intractable diseases database.¹⁸ These findings revealed no similarities in geographic or ethnicity in our prevalence result. Rather, our prevalence was similar to a previous US prevalence of 5.5/100,000 from hospital records review data,¹⁹ Israel incidence of 2.18/1,000,000 from general hospital record review,²⁰ and Japanese prevalence of 2.4/100,000 from enquiries sent to practitioner offices and hospitals.²¹ The variability in prevalence and incidence estimates was attributed to possible methodologic differences in a recent systematic review report.² The highest estimates are noted in register-based studies where cases are defined using administrative codes,^{22–24} thus potentially overestimating the true incidence or prevalence. The lowest estimates are found in studies where cases are defined using classification criteria and biopsy findings,^{25–27} thus potentially underestimating the true incidence/prevalence. Our database using the ICBP registration system may have included patients diagnosed definitely by specialists, although the source was claims data.

Table 2. Comorbidities of prevalent patients with inflammatory myositis according to sub-diagnosis in 2015

Characteristics	JDM (n = 130)	DM (n = 1,101)	PM (n = 869)
Elixhauser comorbidity index, median (IQR)	1 (0, 2)	2 (1, 4)	3 (2, 5)
No. of comorbidities			
0	35 (26.9)	147 (13.4)	85 (9.8)
1	49 (37.7)	177 (16.1)	123 (14.2)
2	17 (13.1)	238 (21.6)	157 (18.1)
≥ 3	29 (22.3)	539 (49.0)	504 (58.0)
Congestive heart failure	1 (0.8)	65 (5.9)	71 (8.2)
Cardiac arrhythmias	1 (0.8)	42 (3.8)	59 (6.8)
Valvular disease	1 (0.8)	4 (0.4)	5 (0.6)
Pulmonary circulation disorders	2 (1.5)	25 (2.3)	30 (3.5)
Peripheral vascular disorders	3 (2.3)	106 (9.6)	87 (10.0)
Hypertension, uncomplicated	5 (3.9)	266 (24.2)	299 (34.4)
Hypertension, complicated	2 (1.5)	31 (2.8)	29 (3.3)
Paralysis	3 (2.3)	20 (1.8)	24 (2.8)
Other neurological disorders	2 (1.5)	37 (3.4)	53 (6.1)
Chronic pulmonary disease	36 (27.7)	372 (33.8)	307 (35.3)
Diabetes, uncomplicated	8 (6.2)	194 (17.6)	190 (21.9)
Diabetes, complicated	2 (1.5)	104 (9.5)	103 (11.9)
Hypothyroidism	-	75 (6.8)	76 (8.8)
Renal failure	1 (0.8)	18 (1.6)	18 (2.1)
Liver disease	15 (11.5)	239 (21.7)	250 (28.8)
Peptic ulcer disease excluding bleeding	15 (11.5)	240 (21.8)	203 (23.4)
AIDS/HIV	1 (0.8)	-	1 (0.1)
Lymphoma	-	13 (1.2)	5 (0.6)
Metastatic cancer	-	-	-
Solid tumor without metastasis	1 (0.8)	130 (11.8)	72 (8.3)
Coagulopathy	4 (3.1)	28 (2.5)	32 (3.7)
Obesity	-	1 (0.1)	-
Weight loss	1 (0.8)	24 (2.2)	15 (1.7)
Fluid and electrolyte disorders	8 (6.2)	81 (7.4)	87 (10.0)
Blood loss anemia	-	1 (0.1)	-
Deficiency anemia	17 (13.1)	233 (21.2)	168 (19.3)
Alcohol abuse	-	12 (1.1)	18 (2.1)
Drug abuse	-	1 (0.1)	1 (0.1)
Psychoses	-	10 (0.9)	11 (1.3)
Depression	6 (4.6)	122 (11.1)	125 (14.4)
Pulmonary disease specified			
Interstitial lung disease	2 (1.5)	240 (21.8)	178 (20.5)
Pneumothorax	-	7 (0.6)	3 (0.4)
Pneumomediastinum	-	9 (0.8)	-

Data are presented as number (%).

JDM = juvenile dermatomyositis, DM = dermatomyositis, PM = polymyositis, IQR = interquartile ranges, AIDS/HIV = acquired immune deficiency syndrome/human immunodeficiency virus.

Several previous studies from the US report a significant increase in the incidence rate of overall IIM over time. The incidence rate increased from the first 11-year period (2.6/million/year) to the second 11-year period (6.5/million/year) from 1947 to 1968 in one study²⁶ and from 2.5 for 1963–1972 to 8.9/million/year for 1973–1982 in another.¹⁹ These trends likely reflect improvements in medical record indexing, increased awareness of physicians with community experience over time, and improved diagnostic tools.² In Korea, the ICBP system for rare incurable diseases was started in 2004. The gradual increase of incidence by 2009 might reflect the effect of promoting the system. Some prevalent patients could be newly registered to this system, which might cause a slight overestimation of incidence. However, there was no increasing tendency over a 10-year period. Further long term follow-up investigation will be needed.

About 88% of patients with IIM had more than one comorbidity. Chronic pulmonary disease was the most common in all types of IIM, with a higher prevalence (20.0%) compared to in RA (11.6%), as reported in Korean claims data.²⁸ We presented the prevalence of various comorbidities using the Elixhauser score, which was more recently developed.¹⁶ This score includes more conditions and is becoming more widely used as recent comparisons suggest that it outperforms the Charlson comorbidity index with use of administrative claims data.²⁹

A key strength of the present study is that it is a large, population-based study using a nationwide database. Furthermore, the national registration system for rare and intractable diseases allowed identification of patients who met the classification criteria for IIM. Because IIM is an extremely rare condition, a larger study population increases the reliability of both incidence and prevalence rates. In addition, an understanding of the prevalence of comorbidities in IIM can facilitate estimation of disease burden and severity.

There were several limitations to this population-based study. First, our case definition may have missed patients with mild symptoms who did not meet classification criteria. Second, we could not conclude whether there was an increase or decrease in trends over time in incidence of IIM because it is very rare disease, and trends could be affected by the starting point of the national registration system for rare diseases in Korea. Going forward, we could update the trends using more long term data. Third, misclassification of sub-diagnoses is possible, and amyopathic DM and inclusion body myositis were not identified in our study due to the lack of specific diagnostic codes. If inclusion body myositis could be added as one of the specific rare diseases in our registration system, we could estimate its prevalence. Fourth, we did not determine the cardiac comorbidities, which have the high mortalities in the patients with IIM, because several cardiac diseases such as myocarditis and pericarditis can be asymptomatic and may not be assigned the appropriate diagnostic codes.

In conclusion, our study observed a slightly lower prevalence and incidence of IIM in Korea compared to recent reports from other countries. More than two-thirds of patients had more than two comorbidities.

SUPPLEMENTARY MATERIAL

Supplementary Fig. 1

Patients' selection flow.

[Click here to view](#)

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