The prevalence of 30 ICD-10 autoimmune diseases in Denmark

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Abstract

Epidemiologic studies of autoimmune diseases have not considered them in the aggregate. The objective was to estimate the prevalence of 30 autoimmune diseases separately and in aggregate according to ICD-10 classification. The lifetime prevalence of the entire population of 5,506,574 persons alive in Denmark on October 31, 2006, was estimated by linking records of all visitors to hospitals and specialty clinics via National Patient Registers from January 1, 1977 through October 31, 2006. The prevalences vary from 0.06/1,000 for Pemphigus to 8.94/1,000 for Type 1 diabetes. Nearly 4% of the population had one or more autoimmune disease. The general conclusion is that autoimmune diseases as an aggregate are common.

Keywords

Introduction

The epidemiology of the autoimmune diseases is underdeveloped because they are usually studied one by one, or in groups based on the organ system that is affected by the autoimmune process [1], although there is at least one systematic review collecting together the results of disparate studies of separate diseases [2]. No population-based studies have considered a range of autoimmune diseases using a single procedure of data collection except an earlier paper by us, which presented data on the prevalence of ICD-8 *or* ICD-10 autoimmune diseases in the

population of Denmark in 2001 [3]. This research note presents new prevalence data through October 31, 2006, with several advantages. The length of the risk period is longer, that is, from 1977 to 2006, instead of from 1977 to 2001. This longer period includes a higher proportion of cases reported in outpatient specialty clinics, which have been a part of the register system only since 1995. This longer risk period and larger number of cases permit the estimation of prevalence strictly according to ICD-10 classification only, which includes refinements as regards autoimmune diseases. This paper defines the autoimmune diseases more strictly than the earlier paper, eliminating diseases and subcategories that might not be considered autoimmune.

Materials and methods

Data from the Danish Civil Registration System [4] were used to identify all persons alive and living in Denmark on October 31, 2006 (5,506,574 persons). The National Hospital Register has collected data on all admissions to Danish Hospitals since 1977. With the exception of a few private clinics performing a small proportion of elective surgical procedures, it includes information from all general hospitals in Denmark. Since 1995, it has included all contacts in emergency rooms and outpatient clinics.

All treatments are free of charge for residents of Denmark. From 1977 to 1993, diagnoses were according to the Danish version of the World Health Organization International Classification of Diseases, eighth revision (ICD8:[5,6]), and from 1994 onwards, according to the International Classification of Diseases 10th edition (ICD10:[7]). People in the study population were classified with a history of an autoimmune disease if they have been admitted to hospital or been in outpatient care in a hospital-based clinic with a diagnosis of the disease in question before October 31, 2006.

The focus here is on the lifetime prevalence of disease, which we define as the proportion in the population in 2006, which has the disease currently or has a history of the disease. The lifetime prevalence for each disease was estimated as the proportion of the persons alive and living in Denmark on October 31, 2006, who were diagnosed with the disease in the period from 1977 to 2006. Individuals with more than one disease occurrences were counted in the numerators of each disease. Since the register began operation in 1977, there is a full lifetime record on all diagnoses given at Danish hospitals and clinics only for persons younger than 30 years on October 31, 2006. Persons older than that will enter the data-set only if a visit is recorded after 1977. This implies an underestimation of the prevalence.

This study was approved by the Danish Data Protection Agency and the Danish National Board of Health.

Results and discussion

Comparison of the categories in Table 1 below (columns 2 and 3) with the categories in Table 1 of the earlier paper [3] reveals that for 15 of the diseases, the categorization is identical and that for other diseases, the categories have been refined to more carefully ensure that the disease is autoimmune in nature. In one disease, the decision was made to expand the ICD-10 category slightly in this analysis (to include M32.9 in Systemic Lupus Erythematosis), and for Rheumatoid Arthritis, the category in this analysis was narrowed in the ICD-8 (including only 712.19, 712.39, and 712.59, instead of the entire category of 712) and expanded in the ICD-10 (including M06 as well as M05). For diabetes, the ICD-8 category of 250 included both Type 1 and Type 2 diabetes. Code 249 was introduced during the ICD-8 period, but the earlier paper used only code 250, thus mixing the two types of diabetes for a subsample of those cases diagnosed during the ICD-8 period.

Prevalence proportions are presented in Table 1 for the combination of ICD-8 and ICD-10, for cases entering treatment from 1977 through 2006 (column 4). These cases have the longest risk period and include the broader ICD-8 classification if they presented for treatment only before 1995. These prevalence proportions include more cases in the numerator, due to the longer follow-up period, and are thus slightly higher than those based only on ICD-10 presented in the rightmost column for cases presenting from 1994 through 2006. The prevalences for the ICD-10 are the most current available in the scientific literature to our knowledge.

The autoimmune diseases range from very rare, such as Pemphigus at 0.06 per 1,000 population, to what might be called infrequent, such as Type 1 Diabetes, at 8.94 per 1,000 population, just under one percent. About half the diseases in Table 1 have prevalences below 0.5 per 1,000, and only four (Thyrotoxicosis, Type 1 Diabetes, Ulcerative Colitis, and Rheumatoid Arthritis) have prevalences higher than 4 per 1,000. The prevalence of any ICD-10 autoimmune disease among those listed is 36.39 per 1,000 or 3.6% (Table 1) and slightly higher for the combination of ICD-8 with ICD-10.

There are a few striking differences between the results summarized in this article and previous reports. They may provide insight with the way that diseases are managed in different countries. For example, earlier studies indicated that the prevalence of autoimmune thyroiditis is about equal to the prevalence of thyrotoxicosis (Graves' disease: 2). The present report gives a prevalence of thyroiditis that is about one tenth that of Graves' disease. We suggest that the difference may indicate that hypothyroidism is frequently diagnosed and treated by non-specialist physicians in Denmark and, therefore, would not be fully accounted in the present study. Vitiligo may be treated less frequently in Denmark than in other countries due to the generally light complexion of Danes. Nevertheless, we may speak generally that autoimmune diseases occur in about 4% of the population.

Acknowledgments

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Table 1

Prevalence of autoimmune diseases in Denmark in 5,506,574 persons living in Denmark on October 31, 2006

Autoimmune disease	Categorization		Prevalence per 1,000	
	ICD8	ICD10	ICD8/ICD10	ICD10 ONLY
Pernicious anemia	281.0	D51.0	0.50	0.37
Autoimmune hemolytic anemia	283.90–91	D59.1	0.13	0.11
Idiopathic thrombocytopenic purpura	446.49	D69.3	0.46	0.45
Thyrotoxicosis	242.00	E05.0	4.51	4.34
Autoimmune thyroiditis	245.03	E06.3	0.58	0.52
Type 1 diabetes	249	E10	9.12	8.94
Primary adrenocortical insufficiency	255.1	E27.1	0.19	0.16
Multiple sclerosis	340	G35	1.89	1.76
Guillain–Barre syndrome	354	G61.0	0.58	0.25
Iridocyclitis	364	H20	2.08	1.85
Crohn's disease	563.01	K50	2.30	2.15
Ulcerative colitis	563.19	K51	4.76	4.34
Autoimmune hepatitis	571.93	K73	0.45	0.33
Primary biliary cirrhosis	571.90	K74.3	0.13	0.12
Celiac disease	269.00	K90.0	0.55	0.49
Pemphigus	694 (×694.05)	L10	0.07	0.06
Pemphigoid	694.05	L12	0.13	0.12
Psoriasis vulgaris	696.09–10, 696.19	L40 (×L40.4)	3.21	2.72
Alopecia areata	704.00	L63	0.31	0.27
Vitiligo	709.01	L80.9	0.22	0.20
Seropositive rheumatoid arthritis	712.19, 712.39, 712.59	M05–M06	5.03	4.64
Juvenile arthritis	712.09	M08	0.73	0.58
Wegener's granulomatosis	446.29	M31.3	0.16	0.15
Dermatopolymyositis	716	M33	0.18	0.14
Polymyalgia rheumatica	446.30–31, 446.39	M31.5–6, M35.3	2.74	2.50
Myasthenia gravis	733.09	G70.0	0.18	0.15
Systemic sclerosis	734.0	M34	0.25	0.23
Systemic lupus erythematosis	734.19	M32.1, M32.9	0.48	0.43
Sjogren's syndrome	734.90	M35.0	0.63	0.59
Ankylosing spondylitis	712.49	M45.9	0.73	0.63
Any of the above diseases	–	–	39.44	36.39