## Accuracy of the ICD-9 Code for Identification of Patients with Wegener's Granulomatosis

*To the Editor:*

The *International Classification of Diseases*, 9th Revision (ICD-9) has been proposed as a valid tool for epidemiological research for some rheumatologic conditions[1,2]. We investigated the accuracy of the ICD-9 code 446.4 for the diagnosis of Wegener's granulomatosis (WG).

The Institutional Review Board of Saint Alexius Medical Center approved the study, and a waiver of informed consent was granted. Saint Alexius Medical Center is a 306-bed hospital, with primary and specialty clinics (including rheumatology, pulmonology, and nephrology). Patients coded for the first time with the ICD-9 code 446.4 were identified using the Health Data Management Reporter software (version 4.7.1000.11; 3M Health Information System Inc., St. Paul, MN, USA).

Between 1996 and 2006, the ICD-9 system was used in 676,851 patients for billing purposes. The code 446.4 was recorded 231 times, of which 44 were for the first time. The electronic records of these 44 patients at the time the code was applied were reviewed, but 21 were excluded because they were missing or incomplete, not allowing further analysis. That left 23 patients in our study.

The clinical characteristics of the patients are presented in Table 1. Twelve (52%) met the modified American College of Rheumatology (ACR) criteria for WG[3] used in a recent drug trial (9 patients had 2 criteria, 2 had 3 criteria, and 1 had 5 criteria). Of these 12 patients, 6 were positive for cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) and 6 were positive for perinuclear ANCA (pANCA). There were 8 patients who had only 1 criterion, of whom 3 were pANCA-positive and were treated with cyclophosphamide. Finally, there were 3 patients who had no criteria for WG; in these patients ANCA testing was ordered to rule out WG; none of these 3 patients received treatment for WG.

Epidemiological studies using large databases can be successful only if the code for a disease is accurate, particularly for rare conditions such as WG. In a study from New Zealand, the *International Classification of Disease* 10th Revision, Australian Modification (a coding system not based on the ICD-9) was used to show evidence for a latitude-dependent incidence gradient of WG[4]. However, the accuracy of this coding system was not analyzed for WG.

In our study, we found in a nonreferral institution for vasculitis that about 65% of the patients classified with the ICD-9 code for WG met the modified ACR criteria or had an ANCA-associated condition. Some limitations need to be acknowledged: our study had a retrospective design, few patients were included, and our study could be biased since about half the patients were excluded because of incomplete records.

The results of our study suggest that the ICD-9 code for WG is of limited value for epidemiological investigations. However, it can be used as an initial screening tool for the identification of patients with WG, if followed by a careful review of the patient records.

*Table 1.* Characteristics of the 23 patients classified as having Wegener's granulomatosis by the ICD-9 coding system. Abnormal chest radiograph means that a patient has nodules, cavities, or fixed infiltrates. Abnormal urinary analysis: presence of red blood cell casts or more than 5 red blood cells per high-power field. Abnormal biopsy implies that the patient has granulomatous inflammation or necrotizing vasculitis.

| Characteristics | |
|---|---|
| Age (SD), yrs | 61 (19) |
| Female, n (%) | 14 (61) |
| White, n (%) | 21 (91) |
| Abnormal chest radiograph, n | 9 |
| Abnormal urinary analysis, n | 7 |
| Abnormal biopsy, n | 7 |
| Nasal discharge or oral ulcers, n | 5 |
| cANCA-positive, n | 9 |
| pANCA-positive, n* | 9 |

* One patient who was pANCA-positive was proteinase 3-positive by direct ELISA. ICD-9: International Classification of Diseases, 9th Revision; cANCA: cytoplasmic antineutrophil cytoplasmic antibodies; pANCA: perinuclear ANCA.

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