Incidence and mortality of relapsing polychondritis in the UK: a population-based cohort study

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Abstract

Objective. Relapsing polychondritis is a rare disease characterized by cartilage inflammation. Our aim was to estimate the incidence, prevalence and mortality of relapsing polychondritis and describe the clinical features of relapsing polychondritis in a large population.

Methods. All participants diagnosed with relapsing polychondritis were sampled from the Clinical Practice Research Datalink. Prevalence and incidence rates for 1990–2012 were estimated. Relative mortality rates were estimated in a time-to-event framework using reference UK life tables. A questionnaire validation study assessed diagnostic accuracy.

Results. There were 117 participants with relapsing polychondritis ever recorded. Fifty (82%) of 61 cases were validated by a physician and unconfirmed cases were excluded. The analysis included 106 participants (42 men, 64 women) diagnosed with relapsing polychondritis. The mean age (range) at diagnosis in men was 55 (range 17–81) years and in women 51 (range 11–79) years. The median interval from first symptom to diagnosis was 1.9 years. The incidence of relapsing polychondritis between 1990 and 2012 was 0.71 (95% CI 0.55, 0.91) per million population per year. There were 19 deaths from any cause. There were 16 observed deaths eligible for survival analysis and 7.4 deaths expected for the UK population of the same age, sex and period. The standardized mortality ratio was 2.16 (95% CI 1.24, 3.51), \( P < 0.01 \). Respiratory disease, cardiac conditions and cancer were the most frequent causes of death.

Conclusion. The incidence of relapsing polychondritis may be lower than previously estimated, and diagnostic misclassification and delay are common. Mortality in relapsing polychondritis is more than twice that of the general population.

Key words: relapsing polychondritis, incidence, prevalence, therapy, mortality, Clinical Practice Research Database.

Introduction

Relapsing polychondritis is a rare autoimmune rheumatic disorder characterized by episodic inflammation of cartilaginous tissue throughout the body [1, 2]. Typical presenting features include chondritis of the nasal bridge, auricular cartilage, ocular inflammation and involvement of the tracheobronchial tree [3, 4]. Destruction of the laryngeal and tracheal cartilage rings may lead to collapse...
of the airways and is associated with a high risk of morbidity and mortality [2–4]. Its rarity often leads to considerable delay in establishing a diagnosis [4].

Relapsing polychondritis is part of the spectrum of systemic autoimmune disorders and may present with similar clinical features to other autoimmune rheumatic diseases such as granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis (Churg–Strauss syndrome) or RA [5–7]. Treatment for relapsing polychondritis is usually with steroids and immunosuppressive drugs, but there are no randomized trials and treatment remains empirical and based on expert opinion [7].

The rarity of relapsing polychondritis makes it difficult to obtain accurate epidemiological data. Most reports are of small case series from specialist centres. These generally describe a female preponderance, and one of the largest studies in the literature (consisting of 200 patients) had a female to male ratio of 1.8:1 [8]. The peak age of onset is between the fourth and fifth decades, but the disease has been described in young children and in the very elderly. All ethnic groups may be affected, but the majority of reported cases in the literature are of white Caucasian descent. Only about 800 cases have been reported in the literature worldwide, and this almost certainly underestimates the frequency of the condition. Given the rarity of relapsing polychondritis and the non-specific clinical manifestations, patients often experience significant delay in diagnosis after the onset of symptoms [1]. There is no standard medical therapy for relapsing polychondritis. Treatment is based on controlling symptoms, and an individual approach is necessary [6]. For patients with mild auricular or nasal symptoms, short-term use of NSAIDs is generally adequate [6]. CSs may be required for more serious manifestations; however, steroid therapy is usually tapered off after acute attacks.

The aim of this study was to investigate the epidemiology of relapsing polychondritis, including the incidence, prevalence and mortality, in a population-based epidemiological study. We also aimed to examine the clinical features of relapsing polychondritis, as well as age at diagnosis, duration and type of symptoms, patterns of treatment, prognosis, survival and causes of death.

Methods

Data source

We conducted a population-based cohort study of patients diagnosed with relapsing polychondritis in the Clinical Practice Research Datalink (CPRD). The CPRD comprises the electronic health records of more than 5 million active patients from over 660 family practices in the UK. The CPRD is designed to capitalize on the registration of >98% of the UK population with National Health Service family practices [9]. The CPRD population is representative of the UK general population (7% of the total UK population) [9, 10]. Data elements include demographics, prescriptions, clinical events and diagnoses, referrals to hospitals, and additional patient information such as height, weight, age, smoking status, alcohol use, immunizations and deaths. Diagnoses recorded in the CPRD have been shown to have high predictive value in validation studies [10]. For entry into the CPRD, practice data must be up to standard for research as set out by the CPRD group. Fully anonymized data are available for analysis. This study received scientific and ethical approval from the CPRD Independent Scientific Advisory Committee for CPRD studies (Ref 13-005).

Study population

A cohort of participants with relapsing polychondritis was identified from CPRD clinical and referral records based on the single Read code for relapsing polychondritis, N33z500. The index date, or diagnosis date, was defined as the date of the first recorded relapsing polychondritis event. In view of the rarity of relapsing polychondritis, all diagnoses ever recorded were included. The start of record for each case was defined as the later of the patient’s registration date at a CPRD practice, or the date the practice joined CPRD and provided up-to-standard data. The end of record for each case was defined as the earliest of the death date, the end of registration date, the last data collection date or 31 January 2012. Cases were classified as incident if they were diagnosed more than 1 year after the start of the CPRD record, and as prevalent if they were diagnosed before the start of the record or up to 1 year after the start of the record.

Validation study

In order to confirm that relapsing polychondritis diagnoses were accurately coded into electronic health records, a validation study was performed. A questionnaire was sent to the family practice associated with each of 117 cases of suspected relapsing polychondritis. The questionnaire was self-completed by a general practitioner at the practice using electronic health records for the individual patient with relapsing polychondritis. The questionnaire included five items: whether the patient had a confirmed diagnosis of relapsing polychondritis; whether a specialist confirmed the diagnosis and specialty involved; whether the patient had a biopsy; the main clinical features; and whether any related autoimmune diseases were present. In order to achieve a high response rate, three mailings of the questionnaire were used.

Statistical analysis

Prevalence rates were estimated using mid-year counts for relapsing polychondritis cases and the CPRD denominator population. These were aggregated by 5-year periods. Incidence rates were estimated using incident relapsing polychondritis cases as the numerator and person-years from the CPRD population as the denominator. CIs were estimated from the Poisson distribution. Age standardization was not performed because the data were sparsely distributed. However, the age distribution of the CPRD population is very similar to that of the UK general population.

The relative mortality of relapsing polychondritis cases was estimated in a time-to-event framework [11]. The start
date was the later of the relapsing polychondritis diagno-
sis date, the start of the CPRD record or the date the
practice joined CPRD and provided up-to-standard data.
The end date was the earliest of the death date, the last
collection date or the end of the CPRD record. Relative
survival was estimated using UK life tables, which pro-
vided estimates of the probability of death by sex, single
year of age and period [12]. Expected deaths by year fol-
lowing relapsing polychondritis diagnosis were estimated
using the strs command in Stata version 12.0 [13].
A standardized mortality ratio was estimated as the ratio
of observed to expected deaths, with a 95% CI estimated
from the Poisson distribution.

Clinical features of the cohort were then determined by
evaluating the frequency distribution of Read codes, Read
terms and type of symptom in the clinical records of study
patients. The National Health Service Browser and CPRD
Medical Dictionary were used to develop a list of Read
codes for relapsing polychondritis related clinical mani-
festations. These included symptoms and conditions af-
flicting the ear, eye, joints, nose, skin and throat. Details of
the codes used are available from the authors. These
codes were used to understand and describe the symp-
toms presented by relapsing polychondritis patients
before and after diagnosis, as well as the time from the
first reported symptom to diagnosis. New onset of coro-
ary heart disease, stroke or diabetes, as common
comorbidities, was additionally evaluated before and
after the diagnosis of relapsing polychondritis. The
British National Formulary [14] and the CPRD Product
Dictionary were used to develop a list of codes for differ-
ent drug therapies, and to subsequently determine any
therapeutic trends in treatment course and treatment
type. For these analyses, only clinical and therapy
events in patients’ up-to-research-standard records
were included.

Results

A cohort of 117 patients was initially identified as being
registered in CPRD with one or more medical (Read)
codes for relapsing polychondritis ever recorded in
CPRD up to 31 December 2012. There were nine cases
with index dates before 1990, and 37 in total with index
dates before 2000. There were 61 completed question-
naires received from the validation study. The overall re-
sponse rate was 52%, and the diagnosis was confirmed
for 50 of 61 (82%) cases. The most frequent reported clin-
ical features were external ear inflammation (70%), arth-
ritis (36%) and nasal inflammation (26%) (Table 1). The
diagnosis was supported by specialist opinion including
rheumatology (60%), dermatology (32%) and ENT (28%).

After excluding 11 unconfirmed cases, there were 106
cases of relapsing polychondritis for further analysis
(Table 2). There were slightly more female patients
(60%) in the cohort than males (40%), with males being
slightly older than females at diagnosis (Table 2). The
mean age at diagnosis for males and females was 55
(range 17–81) and 51 (range 11–79), respectively. There
were 44 prevalent cases diagnosed within 1 year after the
start of the CPRD record, and 62 incident cases diag-
nosed later than 1 year after the start of the CPRD record.

The number of patients with relapsing polychondritis
during each 5-year period increased over time; however,
this increase was accompanied by a concurrent increase
in patients registered in the CPRD (Table 3). The preva-
ience of relapsing polychondritis for the 3-year period
from 2010 to 2012 was 9.0 per million (95% CI 7.6,
10.5), and the overall incidence rate between 1990 and

<table>
<thead>
<tr>
<th>TABLE 1 Results of validation study</th>
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<tr>
<td>Total responses received</td>
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<tr>
<td>Confirmed cases</td>
</tr>
<tr>
<td>Unconfirmed cases</td>
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<tr>
<td>Clinical features</td>
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<tr>
<td>External ear inflammation</td>
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<tr>
<td>Arthritis</td>
</tr>
<tr>
<td>Nasal inflammation</td>
</tr>
<tr>
<td>Eye inflammation</td>
</tr>
<tr>
<td>Skin (vasculitis)</td>
</tr>
<tr>
<td>Major airway disease</td>
</tr>
<tr>
<td>Specialist confirmation</td>
</tr>
<tr>
<td>By rheumatologist</td>
</tr>
<tr>
<td>By dermatologist</td>
</tr>
<tr>
<td>By ENT</td>
</tr>
</tbody>
</table>

Figures are frequency (%) of confirmed cases.

| TABLE 2 Characteristics of 106 participants with relapsing polychondritis from CPRD |
|----------------------------------|----------|
| Number                           | Male: 42 (40) | Female: 64 (60) |
| Age at diagnosis, mean (range)   | 55 (17–81) | 51 (11–79) |
| Period of diagnosis              |          |
| <1990                            | 4 (10)   | 5 (8) |
| 1990–94                          | 3 (7)    | 5 (8) |
| 1995–99                          | 9 (21)   | 8 (13) |
| 2000–04                          | 6 (14)   | 20 (31) |
| 2005–09                          | 13 (31)  | 15 (23) |
| 2010–12                          | 7 (17)   | 11 (17) |
| Prevalent cases*                  | 14 (33)  | 30 (47) |
| Years from registration start to diagnosis, mean (range) | -7.5 (−23.1 to 0) | -8.1 (−23.4 to 1.0) |
| Incident cases*                   | 28 (67)  | 34 (53) |
| Years from registration start to diagnosis, mean (range) | 8.1 (1.1–19.4) | 10.1 (1.3–20.2) |
| Deaths                           | 10 (24)  | 9 (14) |

Figures are frequencies (%) except where stated otherwise.
*Prevalent cases were diagnosed within 1 year of start of patient registration. Incident cases were diagnosed >1 year after start of patient registration. CPRD: Clinical Practice Research Datalink.
2012 was 0.71 per million person-years (95% CI 0.55, 0.91), being slightly higher in women than men (Table 3).

Relative mortality
A total of 19 patients died in the cohort, and the median survival was 3.5 years for male and 3.7 years for female patients who died. The most frequent conditions coded before death were respiratory and cardiac complications, old age and cancer. Eight of the 19 patients who died suffered from depression. One of the patients who died was also diagnosed with granulomatosis with polyangiitis, suggesting that polychondritis may have been the initial presenting diagnosis.

Table 4 shows observed numbers of deaths by 2-year periods following diagnosis. There were 6, 4 and 3 deaths observed in the first three periods of 2 years, while the numbers of deaths expected based on UK mortality rates were 1.7, 1.4 and 1.2. These data suggest that the relative mortality in relapsing polychondritis may be 2–3 times higher than in the general population, particularly in the earlier years following diagnosis. After allowing for age, sex and period, the standardized mortality ratio for the entire period of follow-up was 2.16 (95% CI 1.24, 3.51, \( P < 0.01 \)).

Clinical manifestations/symptoms
The symptoms that patients reported before and after relapsing polychondritis diagnosis are described in Table 5. The most commonly reported symptoms were of the skin, throat, joints, eye and ear. Such symptoms were frequently recorded >2 years before the diagnosis of relapsing polychondritis and were also commonly recorded after relapsing polychondritis diagnosis, with the exception of ear symptoms, which were most frequently recorded in the year before diagnosis. In this study cohort, the median time from first reported symptom to diagnosis was 1.9 years. Four (4%) patients were diagnosed with myelodysplasia.

New diagnoses of comorbidity were observed before diagnosis, including coronary heart disease (two cases), stroke (two cases) and diabetes (seven cases). During the period after the diagnosis of relapsing polychondritis, there were 7 new diagnoses of coronary heart disease, 1 of stroke and 11 of diabetes mellitus.

Treatment
Overall, the most frequently used drugs among this relapsing polychondritis cohort after diagnosis were glucocorticoids (64%) and NSAIDs (45%) (Table 6). The number of patients using these drugs increased steadily...
TABLE 5 Symptoms recorded, and new onset of comorbidity, before and after diagnosis of relapsing polychondritis

| Symptom group | ≥2 years before diagnosis (55) | 1-2 years before diagnosis (58) | 1 year before diagnosis (64) | After diagnosis (99)<a>
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<tbody>
<tr>
<td>Ear</td>
<td>15 (27)</td>
<td>7 (12)</td>
<td>26 (41)</td>
<td>29 (29)</td>
</tr>
<tr>
<td>Eye</td>
<td>30 (55)</td>
<td>10 (17)</td>
<td>16 (25)</td>
<td>39 (39)</td>
</tr>
<tr>
<td>Joints</td>
<td>21 (38)</td>
<td>10 (17)</td>
<td>7 (11)</td>
<td>32 (32)</td>
</tr>
<tr>
<td>Nose</td>
<td>6 (11)</td>
<td>2 (3)</td>
<td>2 (3)</td>
<td>10 (10)</td>
</tr>
<tr>
<td>Skin</td>
<td>22 (40)</td>
<td>7 (12)</td>
<td>17 (27)</td>
<td>29 (29)</td>
</tr>
<tr>
<td>Throat/respiratory</td>
<td>35 (64)</td>
<td>19 (33)</td>
<td>27 (42)</td>
<td>63 (64)</td>
</tr>
<tr>
<td>Related diagnoses</td>
<td>3 (5)</td>
<td>3 (5)</td>
<td>4 (6)</td>
<td>9 (9)</td>
</tr>
<tr>
<td>Myelodysplasia</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (2)</td>
<td>4 (4)</td>
</tr>
<tr>
<td>New onset of comorbidity</td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td>Coronary heart disease</td>
<td>2 (4)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>7 (7)</td>
</tr>
<tr>
<td>Stroke</td>
<td>2 (4)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>1 (1)</td>
</tr>
<tr>
<td>Diabetes mellitus&lt;br&gt;</td>
<td></td>
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Figures are frequency (%). Column totals indicate number of cases with up to standard record eligible for analysis. <a>Seven patient records ended before practice joined the clinical practice research datalink. bIncludes diabetes mellitus medical codes and drug codes.

TABLE 6 Therapy prescribed before and after relapsing polychondritis diagnosis

| Drug class       | ≥2 years before diagnosis (55) | 1-2 years before diagnosis (58) | 1 year before diagnosis (64) | After + including diagnosis (99)<a>
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<tbody>
<tr>
<td>NSAID</td>
<td>33 (60)</td>
<td>19 (33)</td>
<td>22 (34)</td>
<td>45 (45)</td>
</tr>
<tr>
<td>Glucocorticoids</td>
<td>19 (35)</td>
<td>10 (17)</td>
<td>25 (39)</td>
<td>63 (64)</td>
</tr>
<tr>
<td>MTX</td>
<td>2 (4)</td>
<td>2 (3)</td>
<td>6 (9)</td>
<td>24 (24)</td>
</tr>
<tr>
<td>AZA</td>
<td>3 (5)</td>
<td>3 (6)</td>
<td>5 (8)</td>
<td>8 (8)</td>
</tr>
<tr>
<td>HCQ</td>
<td>5 (9)</td>
<td>3 (6)</td>
<td>1 (2)</td>
<td>1 (1)</td>
</tr>
<tr>
<td>Dapsone</td>
<td>1 (2)</td>
<td>1 (2)</td>
<td>1 (2)</td>
<td>1 (1)</td>
</tr>
<tr>
<td>Colchicine</td>
<td>2 (4)</td>
<td>0 (0)</td>
<td>1 (2)</td>
<td>4 (4)</td>
</tr>
<tr>
<td>SSZ</td>
<td>3 (5)</td>
<td>2 (3)</td>
<td>1 (2)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Any of the above</td>
<td>43 (78)</td>
<td>30 (52)</td>
<td>46 (72)</td>
<td>83 (84)</td>
</tr>
</tbody>
</table>

Figures are frequency (%). Column totals indicate number of cases with up to-standard record eligible for analysis. <a>Seven patient records ended before practice joined clinical practice research datalink; CYC, MMF, penicillamine, gold and LEF were prescribed to one patient each.

from 2 years before diagnosis to after diagnosis. MTX (24%) and AZA (13%) were also prescribed to significantly more patients after diagnosis compared with before. Biologic therapeutic agents such as infliximab and etanercept have been introduced into routine clinical practice for the treatment of autoimmune rheumatic diseases. However, in this cohort, there were no reports of biologic agents being used. The total number of prescriptions in the cohort was 6448 or 5.8 prescriptions per person-year. Specific drugs prescribed before and after diagnosis are outlined in Table 6. Diabetes mellitus was numerically more prevalent after the diagnosis of relapsing polychondritis and may have been related to the high usage of glucocorticoids (Tables 5 and 6).

Discussion

Relapsing polychondritis is a rare disease and our data have confirmed this, with the identification of 106 cases recorded during 87 million person-years in a primary care database. The prevalence and annual incidence rates of relapsing polychondritis between 2010 and 2012 were very low. Our data suggest that the mean age at diagnosis is older than previously suggested, with an average age at disease onset between the fourth and fifth decades [8]. The study confirms that both young children and the very elderly may be at risk of developing relapsing polychondritis. There were 19 deaths, and we estimated that the relative mortality in relapsing polychondritis may be 2–3 times higher than in the general population, particularly in the early years following diagnosis.

There is only one previous community-based epidemiological study of relapsing polychondritis [15]. This was a retrospective electronic medical record review of patients in the United States Department of Defense beneficiary population. The study included 50 patients identified using the International Classification of Diseases (ICD)-9 code for relapsing polychondritis, of whom 43 (86%) met
the inclusion criteria, which is similar to our validation data. Their calculated prevalence of relapsing polychondritis was lower compared with our data (4.5 vs 9 per million), while the average disease duration was greater (7.1 vs 3.6 years in the present data). However, the mean time to diagnosis was longer in the previous study compared with the present study data (3.2 vs 1.9 years). The smaller number of patients included in the previous study may have led to less accurate estimates, which may partially explain some of the differences between the two studies. Also, the diagnosis of relapsing polychondritis in the present study was validated by physicians, increasing our confidence in the accuracy of study estimates.

Our data underscore the often significant delay between the onset of the first symptom and diagnosis of relapsing polychondritis. Respiratory symptoms were associated with the longest delay to diagnosis, with a median time of 10.4 years. This is perhaps not surprising, given that respiratory symptoms are among the most common reasons for physician consultations in the UK and given that relapsing polychondritis is an extremely rare disease. Patients with initial symptoms related to the ear and nose were associated with a shorter delay. One possible reason for this variation might be that nasal bridge chondritis and auricular inflammation, especially if recurrent, are sufficiently unusual to trigger a specialist referral.

Our data on mortality are the first to demonstrate that relapsing polychondritis patients are at increased risk of premature mortality, with a standardized mortality ratio of 2.16 compared with the general population. Most previous studies describe the commonest cause of death as being associated with major airways disease and recurrent respiratory infections. Predictors of a poor prognosis in patients younger than 51 years of age include saddle nose deformity, arthritis, laryngotracheal involvement, systemic vasculitis and haematuria [5]. In older patients, only anaemia predicts a worse outcome [5]. As with many other systemic autoimmune diseases, a greater number of organs involved are predictive of a poorer prognosis. Our data raise a question concerning whether a delay in diagnosis might be associated with mortality, but the small number of deaths gave little power to investigate this question. Problems of lead time bias (with patients diagnosed earlier appearing to survive longer) and length bias (with more severe cases being diagnosed more rapidly) may have complicated interpretation of any possible association.

Our data also confirm the association of relapsing polychondritis with myelodysplasia [8]. Most large case series in the literature collect data on clinical features retrospectively, making it difficult to interpret the prognostic impact of individual symptoms. There is now a disease activity index which is a standardized tool for clinical assessment of patients with relapsing polychondritis [16], and should improve data collection.

One of the strengths of the CPRD is that data on prescriptions are carefully recorded. In the UK, about 98% of individuals are registered with a family practice, ensuring that these results are population based. This also provides longitudinal information on therapies used in relapsing polychondritis patients, both before and after diagnosis. As seen in Table 6, small numbers of patients were already prescribed therapies such as MTX, AZA, dapsone, colchicine, HCQ, gold and penicillamine prior to being diagnosed with relapsing polychondritis. Approximately one-third of patients were prescribed CSs, and just over half were on NSAIDs >2 years prior to diagnosis. These data suggest that many of these patients presented with clinical features compatible with other autoimmune rheumatic disorders before a diagnosis of relapsing polychondritis was established. This may also have contributed to the delay in the diagnosis we observed. There was no evidence for a change in the type of drug therapy post diagnosis, with the four most commonly prescribed drugs being CSs, NSAIDs, MTX and AZA. There were no records of biologic agents (i.e. infliximab, etanercept) being prescribed, as these are only used in specialist hospital practices in the UK.

The limitations of this study are similar to those of other CPRD studies of rare diseases [17]. A validation study confirmed over 80% of the relapsing polychondritis cases in the CPRD, which was considered acceptable. The relapsing polychondritis diagnoses recorded in the CPRD were confirmed by rheumatology, dermatology or ENT specialists, according to the practice records. The predictive value of a relapsing polychondritis diagnosis in the CPRD of just over 80% is in the lower part of the range of previous validation studies in CPRD. In their systematic review, Herrett et al. [10] and Jick et al. [18, 19] found that the median proportion of cases confirmed was 89% across all conditions. We acknowledge that we excluded 11 suspected cases based on additional data obtained through the validation study, but lack of equivalent data for participants with no response to the validation study could have led to overestimation of the incidence and prevalence of relapsing polychondritis. However, incidence and prevalence might be underestimated if severe cases are treated exclusively at specialist units, with no diagnosis recorded in primary care records. There was a long interval after diagnosis for many cases, and clinical records may not have been retained at the practice for patients who had died or changed practice in the distant past. However, since relapsing polychondritis is an extremely rare condition with non-specific symptoms, the 80% predictive value is acceptable. Misclassification of relapsing polychondritis diagnoses appeared to be clinically important. Our data revealed that patients were more likely to be misdiagnosed and treated for other rheumatic diseases before being diagnosed with relapsing polychondritis. Another limitation is that data on therapy dosage and duration with individual drugs such as CSs or immunosuppressive agents are difficult to ascertain.

In conclusion, to our knowledge, this is the first study to provide population-based data to estimate the incidence, prevalence, therapy and mortality of patients with relapsing polychondritis in the UK. These data are important for improving clinical recognition of this uncommon but potentially serious condition.
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