

Prevalence of neurodegenerative parkinsonism in an isolated population in south-eastern Moravia, Czech Republic

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Parkinson's disease (PD) and parkinsonism are the most prevalent chronic neurological disorders in the elderly. The prevalence of PD in Europe in the population over 65 years of age is reported as approximately 1.6 %, with a little variation among European countries ranging from 1.1 to 2.2 % [1–6]. It has been suggested that the prevalence of neurodegenerative diseases in a small, isolated European communities might be higher than in the general population [5, 7]. We have observed possible rebound of such a phenomenon, i.e., more frequent referrals of patients with parkinsonism to neurological outpatient clinics, in a small village in south-eastern Moravia (Lipov, 69634 Czech Republic, 1,524 inhabitants according to the 2011 census), where a low regional mobility and a traditional lifestyle are preserved. Subsequently, this population was investigated using a three-stage case ascertainment method that was successfully introduced in a previous neuroepidemiological study in South Tyrol [7]. In the first stage, screening questionnaires with a cover letter were delivered to all of the occupied houses in the village. Residents were

instructed to deliver the completed questionnaire to the local general practitioner's office. In the second stage, those who screened positively for parkinsonism according to the questionnaire were examined by a trained primary care neurologist, and a preliminary diagnosis of parkinsonism was made or eliminated. In the third stage, all respondents in whom parkinsonism was presumed were admitted to the corresponding author's hospital for a detailed examination in the tertiary movement disorders center. In this stage, parkinsonism was diagnosed when the subject met UK-PDBB criteria for the diagnosis of PD [8], i.e. the presence of two of four symptoms (bradykinesia and resting tremor or rigidity or impaired postural stability). Prevalence was calculated by age group as the number of cases relative to the number of village inhabitants.

Out of 1,524 inhabitants of the village, 576 subjects were 50 years of age and older: 346 inhabitants aged 50–64 years, and 230 inhabitants aged 65 years and older. Questionnaires were delivered to all 458 residential houses in the village. Of those, 128 (27.9 %) completed questionnaires were returned to the general practitioner's office. In this first stage, 54 (42.2 %) respondents screened positively for parkinsonism by the questionnaire, experiencing one or more symptoms; 9 returned the completed questionnaire without a return address, so it was not possible to determine their identity. Out of the 54 positive respondents, 45 were invited for the screening examination. The trained primary care neurologist made a diagnosis of possible parkinsonism in 19 subjects (14.8 % of the total respondents). In the third stage, a final neurological examination by a movement disorders specialist confirmed the preliminary diagnosis of parkinsonism in 14 patients (10.9 % of the total respondents). Overall prevalence was 2.4 % (95 % CI 1.2–3.6), the prevalence in the population from 50 to 64 years of age was 1.7 % (95 % CI 0.4–3.0) and

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3.5 % (95 % CI 1.1–5.9) for the population over 65 years of age. Out of the 19 patients whose examination was positive for signs of parkinsonism in the second stage, 2 patients (1.5 %) were finally diagnosed with essential tremor and 3 patients (2.3 %) had no convincing parkinsonian signs. The data of 14 positively diagnosed patients are presented in Table 1. The 14 cases with confirmed parkinsonism were detected among the 128 that responded to the initial survey, who make up only 27.9 % of the total catchment area (and also 22.2 % of the village population older than 50 years). For this reason it can be considered that the prevalence value 2.4 % is probably underestimated and that the actual overall prevalence of parkinsonism in this area will probably be even higher.

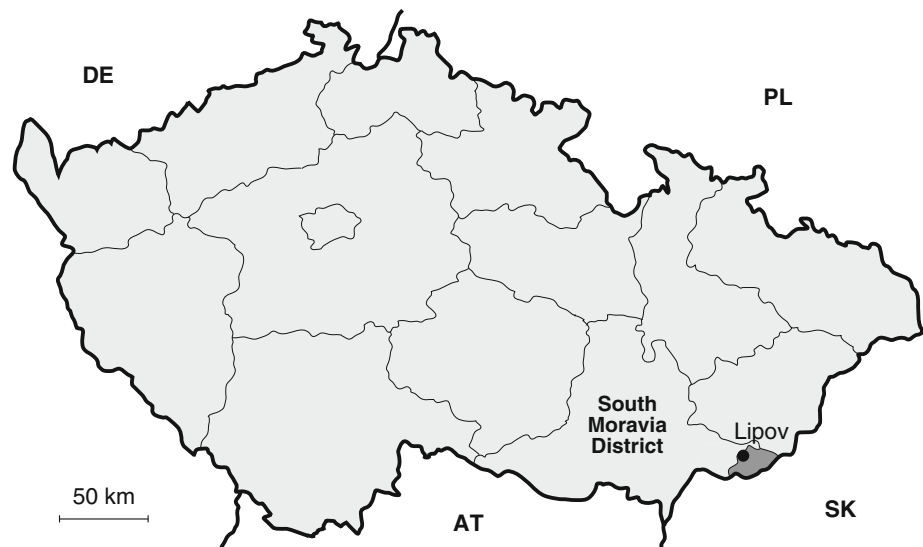
To determine the prevalence of parkinsonism or Parkinson's disease, several door-to-door population surveys were completed and published in the past [1–4]. The door-to-door survey is probably the best way to estimate the prevalence of a disease in a population; however, it is a very costly and labor intensive method. On the other hand, community studies that are based on case detection through physician or hospital visits yield false lower prevalence rates, despite the use of homogeneous diagnostic criteria [6]. For all above-mentioned reasons, we have adopted a three-stage method of case ascertainment for our study, the

methodology which is similar to one that was successfully introduced to investigate the prevalence of parkinsonism and Parkinson's disease in South Tyrol [7]. The prevalence in this study ranged around 1.6 % for Parkinson's disease and 2.2 % for parkinsonism in the population over 65 years. It is important to emphasize here that in our study, we have investigated the incidence of clinical parkinsonism as such. We were not able (in patients in the early stages of the disease) to separate Parkinson's disease subtypes; we were not able to determine in these subjects the direction in which the disorder will develop. In any case, the prevalence in our pilot survey was surprisingly high when compared to previously reported numbers in other parts of Europe [1–5, 7]. It is difficult to explain this difference. Nevertheless, the village we have chosen for our pilot survey is a part of one of the most specific areas in the Central Europe. Hornacko ("Upper Land", Fig. 1) is formed by 10 villages (9,000 inhabitants); it is a small autonomous geographical entity in the White Carpathian mountains (elevation up to 750 m). Its borders are delineated by the catchment area (or basins) of two small watercourses, the Velicka and Kuzelov streams. However, this area is primarily defined ethnographically. The original Slavic population that lived here from the early ninth century was a frequent target of raids during the Hungarian and Turkish invasions in the

Table 1 The clinical data of patients with a confirmed diagnosis of parkinsonism

Pat. nr.	Age	Symptoms at screening	Symptoms at in-ward examination	Treatment
1.	75	Right upper limb rest tremor, head tremor	Right upper limb rest tremor, right upper limb rigidity, head tremor	None
2.	75	Bradykinesia, memory deficit	Bradykinesia, reduced left arm swing, mild cognitive impairment	None
3.	65	Right-sided rigidity	Bilateral rigidity with right-side accentuation	None
4.	65	Right-sided rigidity and bradykinesia	Right-sided rigidity, reduced right arm swing, mild cognitive deficit	None
5.	60	Postural instability, gait disorder	Bradykinesia	None
6.	80	Rest tremor, bilateral rigidity, bradykinesia, postural instability	Left arm rest tremor, bradykinesia, hypomimia	None
7.	61	Bradykinesia, postural instability	Bradykinesia, postural instability	None
8.	53	Rest tremor, postural instability, gait disorder	Hypomimia, rigidity with left-side accentuation, left arm rest tremor	None
9.	59	Rest tremor, rigidity, gait disorder	Rest tremor with right-side accentuation, pull test positivity	None
10.	58	Bilateral hand rest tremor	Bilateral rest and static tremor, reduced right arm swing	None
11.	72	Bilateral hand rest tremor	Right-side rigidity and reduced arm swing, bradykinesia, right arm rest tremor, pull test positivity	None
12.	80	Postural instability, gait disorder	Bilateral rigidity, pull test positivity	None
13.	63	Bradykinesia, gait disorder, intermittent rest tremor	Bilateral rest tremor with right-side accentuation	None
14.	69	Bradykinesia, memory deficit	Bilateral rigidity and bradykinesia, mild cognitive deficit	None

Fig. 1 Map of Czech Republic with highlighted district borders. The dark gray area in the lower right corner is the “Hornacko” region with a highlighted site of the Lipov village. *DE* Germany, *PL* Poland, *AT* Austria, *SK* Slovakia



sixteenth, seventeenth and eighteenth centuries (the last Turkish occupation dates to the late eighteenth century). As a result, this area remained practically uninhabited, and was newly colonized in the mid-nineteenth century, mostly by Lutherans originating mainly in Silesia, western Slovakia, Burgenland, and Styria. These people settled in the remnants of the original Slavic villages and eventually merged to create a homogeneous population, specific and distinct from the surrounding ancient Moravian population in terms of customs, costumes, folk music, and other traditions, including dating and marriage. Marriages outside the area or with potential partners from outside were extremely rare. According to the authorities, to this day the population rarely if ever intermixes with the population outside its 10 villages. This led to rare genetic homogeneity which has been only fractionally altered over the course of the last two centuries, despite all the changes brought by modern life [9, 10]. From this point of view, our study is a unique probe of an ancient population. The question is whether the apparently higher prevalence of parkinsonism in this population (or at least in one of the villages) reflects its original genetic background, or whether it is a result of relative social (and therefore also genetic) isolation and a subsequent high level of in-breeding. Our hypothesis of the influence of genetic factors rather than environmental ones is supported (at least in part) by the observed increased incidence of parkinsonism among blood relatives; and also by the higher incidence of other diseases (such as arterial hypertension) in individual families. To confirm our hypothesis, genetic examinations and further analyses in the families of our 14 probands will be done. To confirm our results in a larger sample, the next step will be to conduct a study using the same methodology and involving the population of all 10 villages.

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