Abstract


Prevalence of Myotonic Dystrophy type 1 in the Västra Götaland Region.

Fredrik Bjerkne, 2014, Institute of Neuroscience and Physiology, Department of Neurology, The Sahlgrenska Academy, University of Gothenburg.

Introduction

Myotonic Dystrophy type 1 (DM1) is the most common inherited muscle disorder in adults with a prevalence in most western countries of approximately 1/ 8000 inhabitants. It’s a multiorgan disorder with a slowly progressive muscle weakness and myotonia i.e. the inability to relax just after muscle contraction. It is classified into four categories; congenital, childhood, adult/classical and late-onset/mild.

Aims

The main purpose of this study was to estimate the prevalence of DM1 in adults in the entire Västra Götaland Region (VGR) as well as in the city of Gothenburg and also in the VGR except Gothenburg, and thus survey possible differences in prevalence between a city and less densely populated areas in the VGR. We also aimed to study if there was a change in diagnostic delay during the four last decades.
Methods

Patients with the diagnosis of DM1 in the VGR were traced by outpatient registers at the Neuromuscular Center (NMC), contacted by regular mail and thereafter telephone interviewed about organ manifestations and debut symptoms. Medical records were examined to obtain detail accuracy.

Results

We detected 230 adult DM1 patients in the VGR which gives a prevalence of 17.8/100 000. The prevalence of DM1 in Gothenburg was 14.1/100 000, which was significantly lower than in the remaining region which was 19.7/100 000. There was no gender difference. We also found that the time gap from symptom onset to diagnose had increased with almost 14 years since the beginning of the 1990’s.

Conclusion

DM1 prevalence in VGR seems to be somewhat higher than elsewhere in the western world. The prevalence result of our survey reiterates the result of previous Swedish study from Örebro County in 1993. We also found a significantly higher prevalence in the less densely populated areas of the region.