



Title	An epidemiological study of motor neuron disease in Hong Kong
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NUS-17 Seronegative myasthenia gravis in Hong Kong Chinese

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Introduction: Acquired myasthenia gravis (MG) is an autoimmune disease due to anti-acetylcholine receptor antibodies (anti-AChR) that damage nicotinic acetylcholine receptors on neuromuscular junctions. Variable proportions of generalized MG patients were reported to have undetectable serum anti-AchR level, designated as seronegative MG (SNMG) but without clear definitions of SNMG. One group reported that antibodies against muscle-specific kinase was the pathogenetic basis of SNMG. The exact pathogenesis of SNMG is not yet certain and genuine frequency is unknown.

Methods: Patients with MG in neurology clinic were studied with clinical, radiological, serological, electrophysiological and histological data reviewed. Patients with initial negative anti-AChR had the assay repeated at least 12 months apart from the first one, together with anti-striational antibody test. Patients with family history of MG had EMG results repeated in details to exclude congenital MG. Patients with repeatedly negative anti-AChR binding assays had serum tested for anti-AChR modulating antibodies (a bioassay using cultured muscle cells) and P/Q type calcium channel antibodies to look for Lambert-Eaton myasthenic syndrome. Only patients with repeatedly negative anti-AchR binding and anti-striational antibodies, negative for anti-AchR modulating and calcium channel antibodies, and with congenital MG excluded were defined as SNMG.

Results: A total 52 MG patients were studied, 21 had pure ocular MG and 31 had generalized MG. Three had turned seropositive upon repeated assay and 1 had positive test for modulating antibodies only. One patient with family history of had congenital MG upon detailed EMG. Only 2 of the 31 generalized MG patients (6.5%) were SNMG. One of the 2 generalized SNMG patient had thymic hyperplasia. No SNMG patients had thymoma. SNMG is more common in pure ocular MG patients.

Conclusion: Generalized SNMG is rare. Stringent criteria are required for diagnosis of SNMG. Thymic hyperplasia, but not thymoma, can occur in SNMG. We postulate that a subgroup of SNMG patients may have low titer of high affinity anti-AchR undetectable by current assays.

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Background: In a previous epidemiological study conducted in 1990s, the incidence and prevalence of motor neuron disease (MND) in Hong Kong Chinese were found to be low compared to the worldwide figures. Moreover, the incidence of MND had been reported to increase steadily over the last few decades in other parts of the world. In this communication, we reported our epidemiological data of MND in the Hong Kong West region of Hong Kong Special Administrative Region of China and compared the results with that of the previous study.

Method: We identified the subjects from the Hospital Authority Database by searching the admission records of Queen Mary Hospital, between 1997 and 2001, using ICD-9 codes of 335. Each retrieved case record was reviewed independently by at least two neurologists. The clinical diagnosis and classification of MND were based on the revised El Escorial criteria.

Results: Of 50 identified subjects, 27 subjects were recruited in the present study. Among which, 22 (81%) were definite MND and 5 (19%) were probable MND. Among definite MND, 15 (68%) were limb onset. The male to female ratio was 2.8:1. All subjects were Chinese. The number of new cases from 1997 to 2001 was 22. Thirteen cases were died in this period, and 14 patients were surviving as of December 31, 2001. Therefore, the annual incidence was 4.4 with an incidence rate of 0.77/100,000/year, the point prevalence at December 31, 2001, was 2.4/100,000, the average annual mortality was 2.6, and the mortality rate was 0.46/100,000/year. The mean survival time was 27.1 months (range: 7 to 65 months, SD 17.9). The mean age of onset was 51.5 years (range: 30 to 77 years, SD 11.7), with a peak observed between 55 and 59 years. Eleven cases (40%) had their disease-onset before 50 years of age.

Conclusion: In this study, the incidence and prevalence of MND among Hong Kong Chinese remained low compared with the worldwide figures. However, comparing to the previous epidemiological data of MND collected between 1989 and 1992, we noticed a trend of increase in the overall incidence (148%) as well as prevalence (152%) over the last decade. Moreover, the incidence of MND among the younger age group was also increased. Our data warrant a territory wide epidemiological study to document the changes in MND epidemiology in Hong Kong over the last decade.

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