The prevalence of multiple sclerosis in Sicily II: Agrigento city

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**SUMMARY**  The prevalence of probable multiple sclerosis in Agrigento city on the south-west coast of Sicily is at least 32 per 100,000. This is likely to be a considerable underestimate of the true prevalence because the study presented particular difficulties in that the city is far from the neurological centres of Palermo, Catania, and Messina. There is no neurological department at either the general or the psychiatric hospital in Agrigento and there was a low awareness of the disease among the doctors in the city. Most of the patients were diagnosed in other centres. Agrigento is a good example of the difficulties of studying multiple sclerosis in a rural city which has no special interest in neurological problems and is far from a neurological centre. Studies in such centres must be pursued with great enthusiasm and over a long period of time, and all available sources of information in the city, medical and lay, and in other cities, must be utilised if a high proportion of the patients is to be found.

Agrigento is a town on the south-west coast of Sicily, famous for its groups of ancient Greek temples. In 1976 the population was 49,979. There is a general hospital in the town but it has no neurological unit and no neurologists attached to it, and there is a large psychiatric hospital. Patients with neurological disorders generally go to Palermo or Catania, or farther afield, to establish the diagnosis. Nearly all the doctors in the town who were interviewed believed that multiple sclerosis (MS) was extremely uncommon; most of them claimed they had not seen a patient with MS resident in Agrigento city.

**Method**

No records could be found of any patients with MS at the general hospital, Ospedale Generale, in Agrigento, but patients' records were not indexed by diagnosis. The husband of one of the nurses at the hospital suffered from MS and this patient was well-documented. Two patients with MS were known to the psychiatrists at the Ospedale Psichiatrica in Agrigento. Records were searched at the psychiatric and general hospitals in Palermo, Catania, Enna, and Messina for any MS patients resident in Agrigento city, and a search was carried out farther afield in Naples, Rome, and Bologna and at the Centro Studi Sclerosi Multipla in Gallarate, Milano. The records of the national health insurance groups were searched for possible MS patients and the clergy of Agrigento were asked if they knew of any patients with symptoms due to MS. The professor of neurology, Professor Agostino Rubino, Palermo, wrote to every doctor in practice in Agrigento asking...
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if they knew of any patients with MS because many of the doctors had studied in Palermo. None of the doctors replied in the affirmative. Lectures were given to the doctors at the general hospital on the diagnosis and epidemiology of MS. All the neurologists in the three university cities in Sicily and those in Naples, Rome, Bologna, and Milan collaborated in the study.

The study in Agrigento can be compared and contrasted with the studies in Enna city,¹ where there was a good neurological clinic, with an excellent records system, and with two neurologists; and in Monreale city,² which is close to the medical school and neurological clinics of Palermo. Patients from Monreale attended hospitals in nearby Palermo because there was no diagnostic hospital in Monreale. Agrigento, in contrast, proved an extremely difficult city in which to carry out an epidemiological study because of the absence of records in the hospitals or doctors’ surgeries, the low awareness of the disease among the doctors, and the distance from a hospital with a neurological unit.

It is likely that more patients with MS resident in Agrigento city will be found in the next few years who had MS on prevalence day and who have been overlooked up to now.

Results

After visiting Agrigento every year for five years and screening the possible MS patients, whose names and addresses were generally found in other cities, 16 patients were considered to have probable MS, seven men and nine women, on prevalence day, 1 January 1975. An additional woman patient was considered to have possible MS and one patient had had retrobulbar neuritis only (Table 1).

Two of the men patients were father and son, otherwise there was no family history among the patients. The first symptoms among the 16 patients with probable MS were paresis in seven and paraesthesia, retrobulbar neuritis, and ataxia in three each respectively.

The average age of onset was 25·6 (21·9 for men and 28·9 for women) and the average age on prevalence day was 41·1 (40·4 men, 41·6 women). Four of the patients had had early onset of symptoms, man no. 3 at the age of 8, man no. 7 at the age of 11, woman no. 6 at the age of 16 and woman no. 7 at the age of 13.

All the patients except woman no. 8, who had had a rapid progressive course, had experienced remissions. One patient committed suicide and three other patients have died since prevalence day. All the patients with probable MS, except one, were born in Agrigento city. Two further long-established patients have been found living in Agrigento now, but they were not resident there on prevalence day and were therefore excluded from the present study.

Sixteen patients with probable MS in Agrigento city in a population of 49 979 is a prevalence of 32·0 per 100 000. If the possible MS patient is included, the prevalence would be 34·0 per 100 000. It is believed that a number of patients with MS resident in Agrigento city, particularly patients with mild symptoms, have not yet been found because of the lack of awareness among the doctors of the symptoms of MS and the delay in making the diagnosis, and because the patients are seldom diagnosed as having MS until they are sufficiently ill to visit the neurological clinics in other cities, usually Palermo. The study in Agrigento will continue and it would appear to be likely that a further report will show a higher prevalence. Although a number of patients in Agrigento have undoubtedly been overlooked, the number of patients found, 16, is not significantly different from the expected number of patients with probable MS based on the prevalence found in Monreale, when the expected number would be 22, or the expected number based on the prevalence in Enna, when it would be 25 (Table 2).

Discussion

The higher prevalence of MS found in Monreale, Enna, and Agrigento cities, in comparison with previous studies in Italy and Sicily, is no doubt due to the deliberate choice of small populations which can be studied in depth. It is very difficult to find all, or nearly all, the diagnosed MS patients even in small populations. In Sicily, few doctors keep medical records and finding patients depends upon the use of many sources of information. Doctors in Sicily believe that MS is very rare and therefore the diagnosis is not likely to come to mind and, when it does, some vague term is often used to avoid naming the disease. The patients in this study were diagnosed in hospitals outside Monreale and not by their home doctor.

The high prevalence of MS found in three cities in Sicily, Monreale, Enna, and Agrigento, confirms the high prevalence found among Italian immigrants to London.³ The slightly lower number of hospitalised MS patients found in immigrants from Spain and Cyprus (Greek and Turkish Cypriots) compared with the expected number at the United Kingdom-born rates, age-corrected, resident in London, suggests that in the south of Italy and Sicily we may also expect to find a slightly lower prevalence than in northern Italy. The studies of MS prevalence in Enna, Monreale, and Agrigento provide evidence that the prevalence in Sicily is between 40 and 60 per
<table>
<thead>
<tr>
<th>No.</th>
<th>Age (years)</th>
<th>Date and first symptom</th>
<th>Remission</th>
<th>Relapse</th>
<th>Remission</th>
<th>Examination</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>56</td>
<td>1940 Unsteady gait</td>
<td>Complete</td>
<td>1948 Blurred vision rt. eye 1958 Weakness legs, could not walk without help 1963 Relapse 1974 Relapse</td>
<td>Yes</td>
<td>Pale discs, nystagmus, ataxia hands, spastic gait, hyperreflexia, Babinski ††</td>
</tr>
<tr>
<td>2</td>
<td>43</td>
<td>1961 Weakness rt. leg</td>
<td>Almost complete</td>
<td>1967 Weakness both legs, blurring of vision, arms ataxic</td>
<td>1964 Relapse</td>
<td>Discs pale, nystagmus, paraparesis R&gt;L, ataxia arms, hyperreflexia R&gt;L, clonus knee and ankle. MS diagnosed Rome, Bologna, etc.</td>
</tr>
<tr>
<td>3</td>
<td>25</td>
<td>1958 (aged 8) Weakness rt. arm and rt. leg, rt. facial paralysis</td>
<td>Complete</td>
<td>1959 Relapse as in 1958. 1964 Bladder symptoms, incontinent, weakness both legs 1976 Severe relapse unable to walk, blurring vision rt. eye</td>
<td>Complete</td>
<td>Vision poor (cannot read), pale discs, nystagmus, ataxia arms and legs L&gt;R, hyperreflexia, spastic legs, knee/ankle clonus Babinski ††, abdominals absent. (Son of man no. 4)</td>
</tr>
<tr>
<td>4</td>
<td>69</td>
<td>1930 Unsteady rt. arm</td>
<td>Complete</td>
<td>1952 Weakness in legs, difficulty in walking, 1966 Relapse, ataxia, difficulty in walking, Still walking however until stroke on 31/8/74</td>
<td>Improved</td>
<td>Nystagmus, ataxia, spastic, paresis, paraesthesia legs, hyperreflexia, Babinski †† (MS diagnosed Palermo, Rome, Padua, etc.). (Father of man no. 3)</td>
</tr>
<tr>
<td>7</td>
<td>13</td>
<td>1972 Weakness rt. face rt. arm, rt. leg</td>
<td>Complete ACTH</td>
<td>1979 Weakness legs, difficulty in walking, speech slurred, weak rt. arm, diplopia, two attacks blurred vision</td>
<td>Improved ACTH</td>
<td>1972 Ospedale Civico, Palermo, rt. Babinski, CSF protein increase, Globulin ++, MS diagnosed 1979 Pale discs, rt. face weak, hyperreflexia R&gt;L, Babinski rt., ataxia rt. arm, ver delayed</td>
</tr>
</tbody>
</table>

Table 1: Probable MS patients resident in Agrigento city, prevalence day, 1 January 1975
<table>
<thead>
<tr>
<th>No.</th>
<th>Age (years)</th>
<th>Date and first symptoms</th>
<th>Remission</th>
<th>Relapse</th>
<th>Remission</th>
<th>Examination</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>36</td>
<td>1965 Burning sensation in legs</td>
<td>Complete</td>
<td>1967 Weakness legs, in bed four months.</td>
<td>Yes</td>
<td>Walks with aids, nystagmus, intention tremor, hyperreflexia, knee/ankle clonus, Babinski (\dagger), abdominals absent.</td>
</tr>
<tr>
<td>7</td>
<td>16</td>
<td>1972 Blurring vision both eyes</td>
<td>Yes</td>
<td>1974 Could not distinguish hot and cold lt. leg. Some months later vertigo, vomiting attacks, weakness lt. leg. Diplopia. Many attacks and remissions up to 1979.</td>
<td>Yes</td>
<td>ACTH 1979/77 Unable to walk, weakness both legs R&gt;L, spastic legs, nystagmus, lt. hand paralysis, ataxic rt. hand, intention tremor pale discs, scanning speech. Pain touch diminished lt. side, vibration sense absent up to arms, abdominals (-), hyperreflexia Babinski (+), July 1980. Deteriorating</td>
</tr>
<tr>
<td>8</td>
<td>37</td>
<td>1974 Weakness in legs Back ache</td>
<td>No</td>
<td>1976 After influenza legs very weak, ataxic and spastic, urgency micturition, incontinent</td>
<td></td>
<td>Hyperreflexia, legs spastic, clonus ankles, Babinski (\dagger), CSF protein increased, Pandy (+), MS diagnosed England, Palermo. Died November 1978</td>
</tr>
</tbody>
</table>

**MS in Agrigento city, Sicily**
100 000, which is of the same order of magnitude as that reported in central and northern Europe (Table 2). Other investigations have shown that MS prevalence increases with subsequent studies, because some patients are inevitably missed in the earlier study. Studies of the prevalence of MS are now required in small cities in other southern European countries such as Greece, Cyprus, and Spain.

The high prevalence in Sicily is in sharp contrast to the low prevalence, 4 per 100 000, in the islands of Malta. Sicily and Malta are therefore at a junction of high and low prevalence in the Mediterranean, and it is in these islands that the genetic and environmental factors responsible for MS may be unravelled.

We thank Professor Giuseppe Grimaldi of Enna, who introduced us to Agrigento physicians; Professor Agostino Rubino, Palermo; Professor Giuseppe Papalia, Messina; Professor Francesco Nicoletti, Catania; Professor Raffaello Gattuso, Catania and Syracuse; Professor Vincenzo Bonavita, Naples; Professor Carlo Cazzullo, Gallarate; Professor Giorgio Macchi, Rome; and Professor Luigi Amaducci, Florence, who assisted us with the studies on MS in Sicily. We also thank all our medical colleagues in Agrigento, especially Professor Mario La Loggia and Dr. Salvatore Bonfiglio; our assistants and interpreters, Ms. Concetta Bisaccia and Dr. Pietro Scolaro; and the MS patients who cooperated in the study.

This study, and other studies on the prevalence of multiple sclerosis in Enna city, in Monreale city, and in the Republic of San Marino, were carried out under a contract with the Commission of the European Economic Community. These studies were under the aegis of the Committee for Medical Research and Public Health (CRM) Specialised Working Group in Epidemiology and Clinical Trials.

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References


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