SHORT REPORT

Ethnic differences in syringomyelia in New Zealand

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Objective: To determine the prevalence of syringomyelia in a defined population in New Zealand and measure the prevalence of syringomyelia in the three main ethnic groups (Maori, Pacific people and Caucasians/others) living in this region.

Methods: A retrospective study of all confirmed cases of syringomyelia diagnosed in residents of northern New Zealand from 1961 to 2003.

Results: In all, syringomyelia was diagnosed in 137 patients. The mean age at onset of symptoms was 27.5 years and mean age at diagnosis was 32.6 years. The incidence of new cases increased from 0.76/100 000 a year between 1962 and 1971 to 4.70/100 000 a year by 1992–2001. The prevalence of syringomyelia in 2003 was 8.2/100 000 people: 5.4/100 000 in Caucasians or others, 15.4/100 000 in Maori and 18.4/100 000 in Pacific people ($\chi^2=37.0$, $p<0.0001$). Syringomyelia was more often associated with an isolated Chiari I malformation in Pacific people (84.4%) as compared with 42.9% of Maori and 38.2% of Caucasians or others ($\chi^2=62.3$, $p<0.0001$).

Conclusion: The prevalence of syringomyelia is higher in northern New Zealand than in studies carried out before the advent of magnetic resonance imaging. The prevalence is particularly high in Maori and Pacific people. The cause of the ethnic differences in the prevalence of syringomyelia identified in this study is unexplained and warrants further investigation.

Studies of syringomyelia conducted before the advent of modern imaging reported prevalences ranging from 3.3 to 8.5/100 000.^{1–3} To our knowledge, there are no prevalence studies of syringomyelia in the English literature after the advent of magnetic resonance imaging. Syringomyelia is believed to affect all ethnic groups. Differences in the prevalence of syringomyelia between ethnic groups have been observed in the US, where syringomyelia was more prevalent in African-Americans than in Caucasians.⁴ In New Zealand, Maori and Pacific children with scoliosis were more likely than Caucasians to have syringomyelia.⁵ In this study, we measured the prevalence of syringomyelia in northern New Zealand and determined if there were ethnic differences in the prevalence of syringomyelia.

METHODS

We retrospectively identified all cases of syringomyelia diagnosed in northern New Zealand (Auckland and Northland) residents from January 1961 to December 2003. The population in this region was 1.3 million in 2001: 12.5% Maori, 11.7% Pacific people and 75.5% Caucasians or others (66% Caucasians with most of the remaining 9.5% being East Asians).⁶ Patients were included if they had symptoms and signs consistent with an intrinsic spinal cord lesion and a spinal cord cavity on MRI, myelography, computed tomography,

surgery or autopsy. Patients with a slit-like cavity were excluded unless there was previous imaging evidence that the cavity had been larger. Patients with syringomyelia secondary to a spinal cord tumour and one patient with inadequate clinical data were excluded.

Patients presenting to Auckland Hospital with syringomyelia between 1961 and 1984 were identified from a previous study.⁷ Those who presented after 1984 were determined by searches of the International Classification of Diseases codes and the neurology and neurosurgery department diagnostic databases. Auckland Hospital provides neurological and neurosurgical services to all residents of northern New Zealand. Patients were excluded if their place of residence was outside the study region.

In New Zealand, the government funds inpatient and outpatient hospital care, but a small minority of patients seek neurosurgical consultations in the private health sector. Since 1985, four neurosurgeons have operated on patients with syringomyelia in private hospitals. A review of the diagnostic database for three of these neurosurgeons identified two additional patients living in the study region. The fourth neurosurgeon did not have a diagnostic database.

Demographic, treatment and outcome data were extracted from hospital notes. The ethnic background of patients is routinely collected in New Zealand hospitals. Maori are the indigenous people of New Zealand. Pacific people comprise all patients who identify themselves as originating from Pacific Island nations, most commonly Samoa, Tonga, the Cook Islands and Niue.

Patients with syringomyelia were classified into subtypes according to the associated pathological abnormalities:

- syringomyelia with an isolated Chiari I malformation;
- syringomyelia with a Chiari malformation and a spinal cord abnormality (spina bifida, diastematomyelia, tethered spinal cord);
- syringomyelia with another structural abnormality of the posterior fossa such as platybasia, but no Chiari or spinal cord malformation;
- syringomyelia with a spinal cord malformation, but normal posterior fossa;
- post-traumatic arachnoiditis;
- other causes of arachnoiditis (infection, subarachnoid haemorrhage);
- idiopathic.

Statistical analysis

Demographic data are presented as mean (SD) and analysed by χ^2 tests. Results were considered significant at the 5% level. Incidence and prevalence figures were calculated from census data.^{6 8 9}

RESULTS

In all, syringomyelia was diagnosed in 137 patients (70 (51.1%) men and 67 (48.9%) women, $p<0.80$) resident in

Auckland and Northland between 1961 and 2003. The diagnosis was confirmed by MRI (110 patients), CT myelography (8 patients), myelography alone (11 patients), or CT (2 patients) and surgery (6 patients). Of them, 68 (49.6%) patients with syringomyelia were Caucasians or other, 28 (20.4%) Maori and 32 (23.4%) Pacific people. The ethnic group was not recorded for 9 (6.6%) patients. For patients for whom the data were available, the mean age at the onset of symptoms was 27.5 (16.8) years (median 28.0; range 0–78 years) and the mean age at diagnosis was 32.6 (17.8) years (median 33.5; range 0–71 years). No difference was found in the age at onset of symptoms or the age of diagnosis among the three ethnic groups.

The year of diagnosis was recorded in 115 (83.9%) patients. The number of new cases diagnosed per year increased over the duration of the study. The incidence per 100 000 population per year was 0.76 from 1962 to 1971, 1.27 from 1972 to 1981, 2.15 from 1982 to 1991 and 4.70 from 1992 to 2001. The prevalence of syringomyelia for those patients alive in 2003 was measured using census data from 2001 (table 1).⁶ In all, 118 patients (86.1%) were alive (55 Caucasians or others, 26 Maori, 29 Pacific people and 8 of unknown ethnic background), 8 (5.8%) were dead and in 11 (8.0%) the status was unknown. The prevalence of syringomyelia in 2003 in all ethnic groups was 8.2/100 000 population.⁶ Syringomyelia was more common in Maori and Pacific people, with prevalences of 18.4/100 000 in Pacific people, 15.4/100 000 in Maori and 5.4/100 000 in Caucasians or others ($\chi^2 = 37.0$, $p < 0.0001$).

The most common subtype was syringomyelia associated with Chiari I malformation (50.4%; table 1). Syringomyelia was more often associated with an isolated Chiari I malformation in Pacific people (27/32, 84.4%) than in Caucasians or others (26/68, 38.2%) and Maori (12/28, 42.9%; $\chi^2 = 62.3$, $p < 0.0001$). The prevalence of syringomyelia associated with Chiari I malformation in 2003 was 1.8/100 000 for Caucasians or others, 7.1/100 000 for Maori and 15.9/100 000 for Pacific people. A trend to a greater frequency of syringomyelia with spinal cord malformations in Caucasians or other over than in Maori or Pacific people was observed ($\chi^2 = 4.64$, $p = 0.098$).

Post-traumatic arachnoiditis was more likely to occur in men (10/12) than in women (2/12), excluding one patient of unknown ethnicity with post-traumatic arachnoiditis. Syringomyelia associated with Chiari I malformation was more likely to occur in Caucasians or other women (18/26) than in Caucasians or other men (8/26). The small numbers of patients in the remaining subtypes make it difficult to comment further on ethnic or sex variations.

DISCUSSION

This study has found that the incidence of syringomyelia increased from 1961 to 2001 and that the prevalence of

syringomyelia was higher in 2003 than in studies conducted previously.^{1–3} The increasing incidence is probably due to two main factors: improved detection of syringomyelia using MRI, which has been available in Auckland only since 1992, and the changing ethnic composition of the New Zealand population.¹⁰ The percentage of Maori and Pacific people in the population has increased over the study period. The population of Pacific people has grown 11 times faster than other ethnic groups, so that a higher rate of syringomyelia in Maori and Pacific people translates into increasing incidence in the overall population.¹¹

The finding that Pacific people and Maori have a higher prevalence of syringomyelia, especially syringomyelia associated with a Chiari I malformation, than Caucasians or other is intriguing. Ethnic variations in the size of the posterior fossa and of cerebrospinal fluid flow in the foramen magnum may underlie these differences.^{12–13} A small posterior fossa volume may facilitate the development of a Chiari malformation and syringomyelia.¹² We could not test this hypothesis in our population, because presurgical scans had not been retained for most patients. Posterior fossa volume has not been studied in Maori and Pacific people, but there are other differences in craniospinal development between Caucasians or other, Maori and Pacific people.^{14–17}

Sex differences were also identified. Caucasians or other women were more likely to have syringomyelia associated with a Chiari I malformation than Caucasians or other men. This difference was not seen in Maori or Pacific people. Sex differences for Chiari I malformation have also been found in 364 patients in the US.¹⁸ Differences in craniospinal development between Caucasian men and women may explain this observation.

Several potential problems were associated with this study. The study was a retrospective review over 42 years. Medical records were incomplete for some patients and the method of diagnosis changed over the study period. The number of people with complete information was, however, sufficiently representative to provide valid results. Some patients managed in the private healthcare system may have been overlooked, but these numbers are small and unlikely to markedly alter the results. Syringomyelia is a rare disorder, but the risk of chance fluctuations changing the incidence of this disorder was minimised by the long period of data collection and the large population base from which patients were selected.

In conclusion, this study has found a higher prevalence of syringomyelia than previous studies. Ethnic differences in the prevalence of syringomyelia have been identified, with a higher prevalence in Maori and Pacific people than in Caucasians or other. Greater numbers of Pacific people, and to a lesser extent Maori, were found to have syringomyelia associated with Chiari I malformations. The cause of these

Table 1 Sex, ethnicity and subclassification of patients with diagnosed syringomyelia from 1961 to 2003

Condition associated with syringomyelia	Caucasians/others			Maori			Pacific people			Unknown	Total
	F	M	Total	F	M	Total	F	M	Total		
Isolated Chiari I malformation	18	8	26 (38.2%)	6	6	12 (42.9%)	15	12	27 (84.4%)	4	69 (50.4%)
Chiari malformation and cord abnormality	6	8	14 (20.6%)	0	3	3 (10.7%)	0	1	1 (3.1%)	1	19 (13.9%)
Other posterior fossa abnormality	1	1	2 (2.9%)	1	1	2 (7.1%)	0	0	0	0	4 (2.9%)
Isolated spinal cord abnormality	0	2	2 (2.9%)	0	0	0	1	0	1 (3.1%)	1	4 (2.9%)
Post-traumatic arachnoiditis	1	7	8 (11.8%)	1	1	2 (7.1%)	0	2	2 (6.3%)	1	13 (9.5%)
Non-traumatic arachnoiditis	3	1	4 (5.9%)	0	2	2 (7.1%)	0	0	0	0	6 (4.4%)
No other abnormality	5	7	12 (17.6%)	4	3	7 (25.0%)	0	1	1 (3.1%)	2	22 (16.1%)
Total	34	34	68	12	16	28	16	16	32	9	137
Prevalence*			5.4			15.4			18.4		8.2

F, female; M, male.

*Prevalence/100 000 population of all patients who were alive in 2003 based on 2001 census data.⁶

ethnic differences is unknown and warrants further investigation.

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