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## **Original Paper**

Nationwide Survey on the Epidemiology of Syringomyelia in Japan

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**Running head**

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## **Abstract**

**Background:** Syringomyelia is a rare disease characterized by abnormal fluid-filled cavities within the spinal cord, and is associated with Chiari malformations, arachnoiditis, or spinal cord tumors. The widespread availability of magnetic resonance imaging (MRI) in Japan has allowed for easy identification of syrinxes. The aim of this study was to survey the clinicoepidemiological characteristics of syringomyelia in Japan.

**Methods:** A 2-stage postal survey was conducted in late 2009. The first survey aimed to estimate the number of patients with syringomyelia, and the second survey aimed to elucidate clinicoepidemiological characteristics. Diagnosis of syringomyelia was based on the findings of MRI or computed tomographic myelography.

**Results:** In the first survey, we received 2133 responses from 2937 randomly selected departments and collected data of 1215 syringomyelia patients (543 men and 672 women). The total response rate for the first survey was 73%. The estimated prevalence of ambulatory syringomyelia patients in Japan was 1.94 per 100 000. In the second survey, the proportion of asymptomatic syringomyelia patients was 22.7%. Chiari type I malformations and idiopathic syringomyelia were the first and second most common etiologies.

**Conclusions:** Our nationwide survey indicated that widespread MRI availability has contributed to the diagnosis of both asymptomatic and idiopathic cases.

## Introduction

Syringomyelia is a heterogeneous disorder characterized by abnormal fluid-filled cavities or cysts within the spinal cord. The etiologies of syringomyelia can include Chiari malformations, arachnoiditis, trauma, and spinal cord tumors [1-3], but the pathophysiology of syrinx development remains enigmatic. Some cases with Chiari Type I malformations manifested asymptomatic syringomyelia [4]. The reported prevalence was 8.2 to 8.4 per 100 000 in Western countries [5, 6]. An epidemiologic survey that collected data from 1243 patients between 1982 and 1991 in Japan showed the predominance of Chiari Type I malformations in syringomyelia, and identified a few cases of spontaneous remission [7]. Surgical treatment for syringomyelia is essential to stop the progression of the disease and further cavity enlargement. However, the previous epidemiologic survey did not determine the prevalence of the disease in the Japanese population [7].

The diagnosis of syringomyelia has been greatly aided by the development and widespread availability of magnetic resonance imaging (MRI) scanners, which have allowed for the relatively easy identification of syrinxes. Japan has the highest number of magnetic resonance imaging (MRI) scanners per capita, with national healthcare insurance coverage allowing universal access to outpatient hospital care. Hence, both symptomatic and asymptomatic syringomyelia patients can be more adequately examined than was possible prior to MRI

facilities becoming widely accessible.

The characteristics of asymptomatic syringomyelia have not been sufficiently investigated.

The aim of this study, therefore, was to estimate the prevalence of syringomyelia in Japan and identify its clinicoepidemiological characteristics by taking advantage of the current widespread availability of MRI facilities.

## **Methods**

We conducted a 2-stage postal survey according to methods described previously [8, 9] in late 2009. The first survey aimed to estimate the number of individuals with syringomyelia, and the second survey aimed to elucidate the clinicoepidemiological characteristics of syringomyelia. We collected data from patients diagnosed with syringomyelia by neuroimaging from the departments of neurosurgery, neurology, orthopedics, and pediatrics. We requested the numbers of male and female ambulatory syringomyelia patients from each department in the past year (August 2008 to July 2009).

In the first survey, we adopted a definition of syringomyelia based on neuroimaging: a central or lateralized syrinx detected on MRI (including syrinxes with septums), or a syrinx detected with computed tomographic myelography in patients who could not undergo MRI because of

metal in the body. The number of patients with syringomyelia in each institution was counted based on this definition. The departments surveyed were randomly selected by stratified sampling from a list of all hospitals with 20 or more beds; the list was obtained from the Ministry of Health and Welfare. Sampling rates were approximately 5%, 10%, 20%, 40%, 80%, and 100% for the stratum of general hospitals with 20 to 99 beds, 100 to 199 beds, 200 to 299 beds, 300 to 399 beds, 400 to 499 beds, and 500+ beds, respectively. Additionally, all university hospitals in Japan were surveyed.

In the second stage of the survey, we requested details of individual patients from each department that had 1 or more syringomyelia patients. The detailed information for each patient was reported based on a retrospective chart review. Epidemiological items included sex, date of birth, time of onset and diagnosis, family history, symptoms and signs, imaging findings, treatment, and clinical course. Symptoms included motor function, sensory disturbance, autonomic failure, cranial nerve disturbance, and skeletal deformity. Motor functions included weakness, muscle atrophy, spasticity, hypotonus, and planter reflex. Autonomic failure included Horner syndrome, anisocoria, dyshidrosis, abnormal nail development, limb hypertrophy, bladder and rectal disturbance, orthostatic hypotension, impotence, and neurogenic arthropathy.

This study was approved by the Institutional Review Board of Hokkaido University.

### *Estimation and statistical analysis*

We estimated the prevalence of syringomyelia based on the results from the first stage of the survey. The estimation was based on the assumption that the responses of the departments were independent of the frequency of patients [8, 10]. Formulas used to estimate the total number of patients, and the 95% confidence intervals are described below.

The point estimation of prevalence was calculated using the following equation, where  $SRT_k$ ,  $RRT_k$ ,  $NS_k$ ,  $n_k$ ,  $N_k$  and  $N_{ki}$  denote the sampling rate, response rate, the number of sampling departments, the total number of departments, the number of responding departments, and the number of departments with  $i$  patients in stratum  $k$ , respectively.

$$\begin{aligned}\hat{\alpha}_k &= \frac{1}{SRT_k RRT_k} \sum_i iN_{ki} \\ &= \frac{1}{\frac{NS_k}{n_k} \frac{N_k}{NS_k}} \sum_i iN_{ki} \\ &= \frac{n_k}{N_k} \sum_i iN_{ki}\end{aligned}$$

### **Results**

In the first survey, we received 2133 responses from 2937 randomly selected departments, and collected data regarding 1215 syringomyelia patients (543 men and 672 women). The total response rate of the first survey was 73%.

Results from the first survey (table 1) showed that the number of syringomyelia patients who were referred to a hospital between August 2008 and July 2009 was 2475 (95% CI: 2051–2899). The estimated prevalence of ambulatory syringomyelia patients in Japan was 1.94 per 100,000. In the second survey, we collected reports from 720 of the 1215 patients from the first survey. The response rate for the second survey was 59%. There were 12 duplicated reports, and thus, we integrated the data reported in them.

Results of the second survey (table 2) described the characteristics of both symptomatic and asymptomatic syringomyelia. The proportion of patients with asymptomatic syringomyelia was 22.7% (161 cases). The mean ages at survey and diagnosis of asymptomatic syringomyelia ( $28.9 \pm 23.3$  and  $24.4 \pm 24.1$  years, respectively) were lower than those of patients with symptomatic syringomyelia ( $40.8 \pm 22.8$  and  $35.3 \pm 22.5$  years, respectively). Asymptomatic syringomyelia tended to be primarily associated with localized cavities. The proportion of syringomyelia cases with a Chiari type I malformation etiology was higher among symptomatic than asymptomatic syringomyelia patients. Conversely, the proportion of cases with idiopathic etiologies was higher in asymptomatic than in symptomatic syringomyelia.

A subset of patients with symptomatic syringomyelia (table 3) included both those who had,

and those who had not undergone surgical treatment. The mean age at onset and diagnosis of patients who had undergone surgical treatment ( $29.4 \pm 21.0$  and  $31.6 \pm 21.5$  years, respectively) was lesser than that of patients who had not received surgical treatment ( $40.1 \pm 22.6$  and  $44.8 \pm 22.3$  years, respectively). There were only 2 cases with a family history of the disease. Approximately 11% of patients in each group experienced an improvement in their symptoms. The most common symptom was sensory disturbance, which was reported in 75.3% of patients with surgical treatment and 68.8% of those without surgical treatment. Motor disturbance was the second most common symptom in each group (59.8% and 51.0%, respectively). Patient histories showed that approximately one-tenth of the patients in each group had previous injuries of the head or spine.

The characteristics of patients in each age group (table 4) showed that the prevalence of idiopathic syringomyelia was higher in adults, particularly in the elderly, than in children.

Figure 1 shows the distributions of patient's ages at the time of survey (fig.-1A), age at diagnosis (fig.-1B), age at surgical treatment (fig.-1C), and year of diagnosis (fig.-1D). The distribution of ages at survey consisted of 2 peaks, at 10 to 20 years of age, and at 60 to 70 years of age. The distribution of age at diagnosis showed a higher proportion of 0- to 20-year-olds. Finally, the distribution of diagnosis year showed an acute increment in the number of

cases diagnosed in more recent years.

## **Discussion**

This study revealed the prevalence (1.94 per 100 000) and characteristics of ambulatory syringomyelia patients in Japan. Among these patients, the prevalence of asymptomatic syringomyelia was 22.6%, and that of idiopathic syringomyelia was 15.8% according to the second survey.

The prevalence of syringomyelia in this survey is lower than that in previous studies that used different methods for estimation [5, 6]. Estimation of prevalence in this survey was based on patients who were referred to a hospital for evaluation or treatment. Therefore, the data from patients whose syringomyelia was stable and who had discontinued their ambulatory care were not collected in this study. It is noteworthy that the early detection of syringomyelia by MRI can allow for early interventions, including surgery. Early diagnosis and intervention is more likely to lead to a positive outcome, and may therefore reduce the number of patients requiring ambulatory care. The lower number of patients diagnosed in the years preceding 2005 (fig.1-D) is consistent with our speculation. However, these results show the characteristics of ambulatory care among syringomyelia patients.

The etiology of syringomyelia can include Chiari malformation, trauma, arachnoiditis, and idiopathic origin, among other causes. In our study, Chiari malformations, including both type I and II, were the most common cause in both children and adults, and this finding is consistent with those of previous studies [7, 11]. In particular, Chiari malformation is more frequent in children than in adults. These results may be associated with the widespread availability of MRI, which contributes to early diagnoses in cases of syringomyelia caused by Chiari malformation. Interestingly, idiopathic syringomyelia was the second most common cause according to our survey. Bogdanov et al. suggested that idiopathic syringomyelia is associated with a small posterior fossa with a narrow cerebrospinal fluid (CSF) space as well as with Chiari I malformation [12]. It is possible that some of the cases of idiopathic syringomyelia in our survey may be attributable to a small posterior fossa. Holly et al. described slit-like syrinx cavities characterized by remnants of the central canal and an asymptomatic clinical course [13]. Therefore, idiopathic syringomyelia has several potential causes, including congenital remnants of the central canal and acquired dilations by a small posterior fossa. Hida et al. reported an association between syringomyelia with Chiari I malformation and birth injuries [14]. In this study, patients with problem at delivery accounted for 2.0% of symptomatic syringomyelia cases, but it had a higher unknown/missing proportion in the past history. Nakamura et al. discuss 2 types of idiopathic syringomyelia: localized and extended. Localized syringomyelia is associated with congenital

enlargement of the central canal of the spinal cord and can be managed conservatively [15].

Actually, most of the patients with idiopathic cases in our study did not undergo surgical treatment. Idiopathic syringomyelia might be less progressive than syringomyelia with other causes.

Asymptomatic syringomyelia comprised 22.7% of all syringomyelia cases in our second survey. Prior to this survey, the proportion of asymptomatic syringomyelia cases was unknown. Cases of a few patients with asymptomatic syringomyelia caused by a brain tumor of the posterior fossa have been previously reported [16-18]. The infrequency of asymptomatic syringomyelia seems inconsistent with our survey results. There are 2 possible explanations for the relatively high proportion of asymptomatic syringomyelia in our survey. Firstly, the symptoms of patients who did not complain because of their age were underestimated. Secondly, the availability of MRI in Japan has resulted in an increase in the number of incidental diagnoses of asymptomatic syringomyelia including slit-like syrinx cavities.

Resolution of syringomyelia without surgical treatment was observed in 17 patients (3.2% of symptomatic patients) in our second survey. Spontaneous resolution of syringomyelia has recently been found to be more common than previously thought [19]. The mechanisms

involved in the development and spontaneous resolution of syringomyelia are unclear despite multiple hypotheses [20]. The number of patients with spontaneous resolution may be underestimated because cases of asymptomatic syringomyelia patients who had not sought consultation were not evaluated in our survey.

Symptoms of syringomyelia include pain, sensory disturbance, and amyotrophy. Bogdanov et al. reported that 90% of patients had unilateral or bilateral sensory disturbances, while 79% of patients experienced weakness or wasting of the upper limbs [21].

Familial syringomyelia cases with autosomal dominant or recessive inheritance have been reported [22, 23]. Chatel et al. suggested that the incidence of familial syringomyelia is approximately 2% [24]. However, a large-scale survey has not yet been conducted to determine the proportion of familial cases. In our study, familial syringomyelia comprised only 2 cases (0.6%) of patients with a reported family history. Although a potentially large number of patients who have been lost to follow-up affect the accuracy of the proportion of syringomyelia, familial syringomyelia cases are extremely rare.

This study has several limitations. Firstly, the prevalence of syringomyelia reported in this study was calculated using the estimated number of ambulatory patients. Cases of patients

who did not receive ambulatory care in the past year were not evaluated. Therefore, the potential number of syringomyelia patients may be larger than that reported in this study.

Secondly, this cross-sectional survey could not evaluate the entire clinical course of syringomyelia. The disease progression from asymptomatic to symptomatic is particularly unclear. The clinical course of idiopathic cases is also unclear. Further investigation is required to determine the most appropriate evaluations and treatments for these patients.

Thirdly, the response rates in this study were 73% and 59% in the first and second stage surveys, respectively. Characteristics of patients whose cases were not reported in the second survey are unknown. The effect of this selection bias on our results is also unknown.

Finally, the definition of syringomyelia associated with spinal cord tumor has been changing, and peritumoral cysts have been differentiated from other distinct forms of syringomyelia. In this study, syringomyelia associated with spinal cord tumor was regarded as merely 1 type of syringomyelia.

Taken together, the findings of our survey can contribute to the development of healthcare services for syringomyelia patients. Knowledge of the characteristics of asymptomatic and symptomatic syringomyelia patients without surgical treatment can be useful for the optimization of those services. Further evaluations of the potential number of non-ambulatory

syringomyelia patients should be performed to estimate the precise prevalence of syringomyelia.

In conclusion, we have investigated the epidemiology of syringomyelia in Japan.

Asymptomatic and idiopathic syringomyelia cases are more common than was previously believed. The widespread availability of MRI scanners has potentially contributed to the early diagnosis of these cases.

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Dr. Sakushima reports no disclosures.

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**Table 1.** Summary of data collected in the first stage of the survey

Types of Departments	Types of Hospitals and Beds	Total No. of Departments	Sampling Rate (%)	No. of Surveyed Departments	No. of Departments that Responded	Response Rate (%)	No. of Reported Patients	No. of Estimated Patients
Neurosurgery	General hospitals with ≤99 beds	710	5%	35	22	63%	0	0
	General hospitals with 100–199beds	528	10%	52	27	52%	7	137
	General hospitals with 200–299 beds	298	20%	59	37	63%	26	209
	General hospitals with 300–399 beds	296	40%	119	73	61%	23	93
	General hospitals with 400–499 beds	167	80%	133	94	71%	40	71
	General hospitals with ≥500 beds	216	100%	216	147	68%	133	195
	University hospitals	113	100%	113	94	83%	267	321
Subtotal		2328		727	494	68%	496	1027
Neurology	General hospitals with ≤99 beds	506	5%	25	13	52%	0	0
	General hospitals with 100–199beds	335	10%	34	18	53%	3	56
	General hospitals with 200–299 beds	170	20%	34	27	79%	6	38
	General hospitals with 300–399 beds	170	40%	68	38	56%	7	31
	General hospitals with 400–499 beds	91	100%	91	59	65%	21	32
	General hospitals with ≥500 beds	93	100%	93	60	65%	25	39
	University hospitals	118	100%	118	103	87%	53	61
Subtotal		1483		463	318	69%	115	257
Orthopedics	General hospitals with ≤99 beds	2278	5%	114	66	58%	4	138
	General hospitals with 100–199beds	1047	10%	105	70	67%	10	150
	General hospitals with 200–299 beds	436	20%	87	63	72%	10	69
	General hospitals with 300–399 beds	362	40%	145	110	76%	48	158
	General hospitals with 400–499 beds	190	80%	152	107	70%	20	36
	General hospitals with ≥500 beds	228	100%	228	178	78%	120	154
	University hospitals	118	100%	118	98	83%	300	361
Subtotal		4659		949	692	73%	512	1065
Pediatrics	General hospitals with ≤99 beds	1069	5%	54	32	59%	0	0
	General hospitals with 100–199beds	613	10%	62	41	66%	0	0
	General hospitals with 200–299 beds	356	20%	71	49	69%	0	0
	General hospitals with 300–399 beds	339	40%	136	105	77%	7	23
	General hospitals with 400–499 beds	184	80%	147	120	82%	11	17
	General hospitals with ≥500 beds	214	100%	214	183	86%	58	68
	University hospitals	114	100%	114	99	87%	16	18
Subtotal		2889		798	629	79%	92	126
Total		11359	26%	2937	2133	73%	1215	2475

**Table 2.** Demographics of patients in the second stage of the survey

	Symptomatic (N = 543)	Asymptomatic (N = 161)	Total (N = 708*)	Missing
Age at Survey (Mean ± SD)	40.8 ± 22.8	28.9 ± 23.3	38.0 ± 23.5	35
Age at Diagnosis (Mean ± SD)	35.3 ± 22.5	24.4 ± 24.1	32.7 ± 23.4	66
Sex (%)				
Male	41.6	44.1	42.1	1
Female	57.3	53.4	56.5	3
Missing	1.1	2.5	1.4	0
Morphology (%)				
Asymmetry	31.3	8.1	25.8	0
Symmetry	58.9	83.2	64.4	2
Missing	9.8	8.7	9.7	2
Distribution (%)				
Syringobulbia				
Bulbus only	1.5	0.6	1.3	0
Bulbus and Spinal Cord	5.7	1.2	4.8	1
Syringomyelia				
Cervical cord only	18.6	32.9	21.8	0
Thoracic cord only	7.9	8.7	8.2	1
Lumbosacral cord only	0.9	9.9	3.1	1
Cervical–Thoracic	49.4	27.3	44.1	0
Thoracic–Lumbosacral	2.6	4.3	3.0	0
Cervical–Lumbosacral	4.6	4.3	4.5	0
Missing	8.8	10.6	9.3	1
Etiology (%)				
Chiari type I	53.6	30.4	48.0	0
Chiari type II	4.4	20.5	8.1	0
Bone anomaly	1.1	0.6	1.0	0
Arachnoiditis	5.7	2.5	4.9	0
Trauma	9.6	0.6	7.5	0
Spinal cord tumor	5.2	5.6	5.2	0
Idiopathic	12.9	24.8	15.7	1
Other	6.1	13.0	7.9	2
Suspected two or more	1.1	1.2	1.1	0
Missing	0.4	0.6	0.6	1

\*Four patients who did not report on the existence of symptoms were excluded.

**Table 3.** Demographics, clinical history, and manifestations of symptomatic patients

		Surgical Treatment		Total	Missing
		Yes	No		
Number of Case		376	157	543	10
Age at onset (Mean ± SD)		29.4 ± 21.0	40.1 ± 22.6	32.3 ± 22.0	
Age at diagnosis (Mean ± SD)		31.6 ± 21.5	44.8 ± 22.3	35.3 ± 22.5	
Age at surgery (Mean ± SD)		32.6 ± 21.0			
Family history (%)	Yes	0.3	0.6	0.4	0
	No	64.4	59.9	62.2	2
	Unknown/Missing	31.1	35.0	32.4	
Course of symptoms after initial diagnosis (%)					
Worsen		51.1	22.3	42.2	2
Unchanged		26.3	56.7	35.5	5
Improved		11.2	10.8	10.9	0
Stop after progression		4.8	5.7	5.0	0
Missing		6.6	4.5	6.4	
Symptoms (%)					
Motor	Yes	59.8	51.0	57.5	7
	No	37.8	45.9	39.4	0
	Unknown/Missing	2.4	3.2	3.1	
Sensory	Yes	75.3	68.8	72.7	4
	No	19.9	21.0	19.9	0
	Unknown/Missing	4.8	10.2	7.4	
Autonomic	Yes	20.7	19.1	19.9	0
	No	65.2	65.6	64.6	3
	Unknown/Missing	14.1	15.3	15.5	
Cranial nerves	Yes	10.1	7.0	9.2	1
	No	83.2	80.9	81.4	2
	Unknown/Missing	6.6	12.1	9.4	
Skeletal deformity	Yes	31.4	22.9	29.3	5
	No	64.9	75.2	67.4	4
	Missing	3.7	1.9	3.3	
Past History (%)					
CNS infections	Yes	3.5	3.8	3.7	1
	No	80.6	74.5	78.3	5
	Unknown/Missing	16.0	21.7	18.0	4
Injuries of head or spine	Yes	11.4	10.2	10.9	0
	No	76.3	75.8	75.7	5
	Unknown/Missing	12.2	14.0	13.4	
Surgery of head or spine	Yes	13.8	12.1	13.4	2
	No	77.4	77.7	77.0	5
	Unknown/Missing	8.8	10.2	9.6	
Problems at delivery	Yes	2.1	1.3	2.0	1
	No	66.2	59.9	63.9	4
	Unknown/Missing	31.6	38.9	34.1	

**Table 4.** Summary of characteristics of patients according to age group

Age	Female (%)	Asymptomatic (%)	Etiology		Localized cavity (%)
			1st 2nd 3rd	%	
<10	51.11	40.9	Chiari type I	40.6	36.9
			Chiari type II	34.8	
			Other	14.5	
10–19	66.07	23.9	Chiari type I	78.8	36.2
			Idiopathic	6.2	
			Other	5.3	
20–29	52.63	14.0	Chiari type I	47.4	47.1
			Idiopathic	22.8	
			Trauma	14.0	
30–39	46.34	20.5	Chiari type I	49.4	39.0
			Idiopathic	17.3	
			Trauma	14.8	
40–49	55.17	15.3	Chiari type I	55.9	27.8
			Idiopathic	18.6	
			Spinal cord tumor	10.2	
50–59	69.74	14.5	Chiari type I	42.1	40.6
			Idiopathic	23.7	
			Spinal cord tumor	10.5	
60–69	54.55	11.5	Chiari type I	28.2	42.9
			Idiopathic	24.4	
			Trauma	16.7	
>70	66.67	24.3	Idiopathic	37.8	40.0
			Chiari type I	27.0	
			Arachnoiditis	13.5	

**Figure legend**

Figure 1.

(A) Histogram showing age distribution of patients at time of survey.

(B) Histogram showing age distribution of patients at diagnosis.

(C) Histogram showing age distribution at time of surgery.

(D) Histogram showing the diagnosis by year.

**Figure 1.**

