

## Original Research Article

# Factors predicting the occurrence of syringomyelia in patients with Chiari I malformations

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### ABSTRACT

**Background:** Chiari I malformations are defined as the downward herniation of cerebellar tonsils more than 5 mm through the foramen magnum. Syringomyelia is a common association of Chiari I malformation.

**Methods:** This purpose of this study was to determine the various factors predicting the occurrence of syringomyelia in patients with Chiari I malformations. This retrospective study was conducted in Government medical college, Thrissur, Kerala, India. 27 patients admitted with diagnosis of symptomatic Chiari I malformations during the period of January 2015 to January 2019 were selected for the study.

**Results:** The mean age was 22.8 years and syringomyelia was more common in patients older than 10 years (p value 0.005). Females were 17(63%) and males were 10(37%) but there no difference in occurrence of syringomyelia among both sexes(p value is 0.16). Syringomyelia was present in 17(63%) cases. Association between various factors and syringomyelia measured by calculating P value which was 0.005 for age >10 years, 0.16 for sex, 0.093 for duration of symptoms >12 months, 0.05 for tonsillar descend >10mm and 0.097 for hypertension. The p value for the association of duration of symptoms >12months and extend of tonsillar descend is 0.001 Mean duration of symptoms (onset of first symptom to the time of presentation) was 16.3 months and syringomyelia was more common in patients with duration of symptoms more than 12 months but this difference was statistically not significant (p value 0.093). Mean tonsillar descend from the level of foramen magnum was 11.3 mms and syringomyelia was more common in patients with tonsillar herniation more than 10mm (p value 0.05). There is a statistically significant relationship between duration of symptoms more than 12 months and tonsillar descend more than 10mm(p value 0.001). There is no significant association between hypertension and occurrence of syringomyelia in patients with Chiari I malformation.

**Conclusions:** Incidence of Chiari I Malformations is more among adults and it is slightly higher in females. Syringomyelia is a common association of Chiari I Malformations. The occurrence of syringomyelia in patients with Chiari I Malformations associated with increasing age of patients and extend of tonsillar herniation. There is no statistically significant association between syringomyelia and duration of symptoms or hypertension.

**Keywords:** Chiari I malformations, Syringomyelia, Tonsillar herniation

### INTRODUCTION

In 1883, John Cleland described about hindbrain herniation in a patient with myelodysplasia and after this report Hans Chiari described various types of hindbrain

herniations in postmortem specimens.<sup>1</sup> Chiari I malformation is defined as the downward herniation of cerebellar tonsils more than 5 mm through the foramen magnum (FM).<sup>2</sup> Chiari I malformation also known as adult Chiari malformation because it usually become symptomatic in adulthood. Syringomyelia is a common association of Chiari I malformation.<sup>3-5</sup> The purpose of this study is to determine the association between various factors associated with syringomyelia in patients with Chiari I malformations.

**METHODS**

The aim of this study was the various factors predicting the occurrence of syringomyelia in patients with Chiari I malformations.

This was a retrospective cross-sectional study, conducted in the department of Neurosurgery, Government medical college, Thrissur, Kerala, India. The patients admitted with diagnosis of Chiari I malformations during the period of January 2015 to January 2019 were included for the study. Consent was obtained from all patients. Data collected retrospectively from medical records. Demographic data, duration of symptoms, comorbidities, tonsillar descend and syringomyelia were documented. The downward herniation of cerebellar tonsils more than 5 mm through the foramen magnum in the MRI brain and craniovertebral junction was taken as the criteria for diagnosing Chiari I malformations. A total of 27 diagnosed cases of Chiari I malformations during this study period were included in this study. Patients with vermian or brainstem descend and those who were not willing to participate in the study were excluded from the study. Statistical analysis was done using SPSS software. Level of significance determined by calculating p value (<0.05).

**RESULTS**

The mean age was 22.8 years. Females were 17 (63%) and males were 10 (37%). Mean duration of symptoms (onset of first symptom to the time of presentation) was 16.3 months. Mean tonsillar descend from the level of foramen magnum was 11.3 mms. Syringomyelia was present in 17 (63%) cases (Table 1).

**Table 1. Variables and their mean and standard deviation.**

Age	Mean: 22.8 years SD: 13.38
Sex	Male: 10(37%) Female: 17(63%)
Duration of symptoms	Mean: 16.3 months SD: 11.51
Tonsillar descend	Mean: 11.3 mm SD: 4.35
Syringomyelia	Present in 17 (63%)

Statistical analysis using SPSS software was done to determine the association between various factors associated with syringomyelia in patients with Chiari I Malformation (Table 2). Association between various factors and syringomyelia measured by calculating P value which was 0.005 for age >10 years, 0.16 for sex, 0.093 for duration of symptoms >12 months, 0.05 for tonsillar descend >10 mm and 0.097 for hypertension. The p value for the association of duration of symptoms >12 months and extend of tonsillar descend is 0.001.

**Table 2: Association between different variables of Chiari I malformations and syringomelia.**

Association between variables (N=27)	p-value
Age >10 years and syringomyelia	0.005
Sex and syringomyelia	0.16
Duration of symptoms >12 months and syringomyelia	0.093
Tonsillar descend >10 mm and syringomyelia	0.05
Hypertension and syringomyelia	0.097
Duration of symptoms >12 months and extend of tonsillar descend	0.001

**DISCUSSION**

Chiari I malformation usually become symptomatic in adulthood. Mean age of presentation in this study was 22.8 years. Occurrence syringomyelia is more among adults. Authors found that it was more common in patients older than 10 years (p value 0.005). Chiari I malformation is more common among females male to female ratio of approximately 2:3. 8 Females are commonly affected in our study also (63%) and on analysis it was found that there no difference in occurrence of syringomyelia among both sexes (p value is 0.16).<sup>6,7</sup>

The radiological definition of Chiari I malformation is tonsillar herniation of at least 5 mm below the foramen magnum according to most of the studies and w Authors e also considered 5 mm as the cut off for the diagnosis. In our study, mean value of tonsillar descend was 11.3 mm. Authors found a statistically significant relationship between the occurrence of syringomyelia and extend of tonsillar herniation. Syringomyelia was more common in patients with tonsillar herniation more than 10 mm (p value 0.05). Authors found that syringomyelia was more common in patients with duration of symptoms more than 12 months but this difference was statistically not significant (p value 0.093).<sup>6,9</sup>

Occurrence of syringomyelia among patients with Chiari I malformation was 30%-70% according to literature.<sup>3-6</sup> Syringomyelia was present in 63% of cases in this study. Tonsillar herniation cannot predict the severity and type of symptoms but patients with tonsillar herniations greater than 12 mm were usually symptomatic and

approximately 30% of patients with tonsils herniating 5-10 mm below the foramen magnum were asymptomatic.<sup>10-14</sup> Authors found that there is a statistically significant relationship between duration of symptoms more than 12 months and tonsillar descend more than 10 mm (p value 0.001).

Chiari I malformation should be considered in patients with resistant hypertension and usually it will resolve after foramen magnum decompression.<sup>15-17</sup> Authors statistically analysed the association between hypertension and occurrence of syringomyelia in patients with Chiari I malformation but it was found to be statistically not significant (0.097).

## CONCLUSION

Incidence of Chiari I Malformations is more among adults and it is slightly higher in females. Syringomyelia is a common association of Chiari I Malformations. The occurrence of syringomyelia in patients with Chiari I Malformations associated with increasing age of patients and extend of tonsillar herniation. There is no statistically significant association between syringomyelia and duration of symptoms or hypertension.

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