OPEN ACCESS

ORIGINAL ARTICLE

The Impact of Syrinx on Clinical Outcomes in Arnold-Chiari Malformation: A Retrospective Study

Pir Tufail Ahmad, Mohammad Nasir, Rizwan Ali, Imran Khan
Department of Neurosurgery, MTI-Hayatabad Medical Complex, Peshawar

ABSTRACT

Objective: Arnold-Chiari malformation is congenital or acquired anomaly of the hindbrain that leads to crowding of the cerebellum and brainstem in the posterior cranial fossa. There is an interplay between syringomyelia and Arnold-Chiari malformation. The objective of this study was to analyse the incidence trends, morphological patterns, and clinical outcomes of Arnold-Chiari malformation with and without syringomyelia, focusing on patients treated at Hayatabad Medical Complex, Peshawar.

Methodology: This retrospective study was conducted on 150 patients from January 2023 to December 2023. Patients were divided into two groups: 75 with syringomyelia and 75 without syringomyelia. Key variables included tonsillar herniation, syrinx length, and surgical outcomes. Statistical analyses were done using t-test and chi-square test to assess relationship between variables, with a significance threshold of $p<0.05$.

Results: Patients with syringomyelia exhibited significantly higher tonsillar herniation (mean: 10.8 ± 2.3 mm) compared to those without syringomyelia (mean: 7.4 ± 1.9 mm) with $p<0.001$. Syrinx presence was significantly associated with surgical outcomes ($p<0.05$). Among patients with syrinx, those with improved surgical outcomes had a mean syrinx length of 18.6 ± 7.5 mm, while stable or unimproved cases exhibited larger syrinx lengths. A significant difference in surgical outcomes was noted between the groups, emphasizing the clinical impact of syrinx presence.

Conclusion: A strong association was found between syrinx presence and clinical outcomes in Arnold-Chiari malformation. Early diagnosis and timely intervention are critical for improved patient outcomes.

Keywords: Arnold-Chiari malformation, Surgical outcomes, Syringomyelia, Tonsillar herniation

Authors' Contribution:
[1,2]Conception; Literature research; manuscript design and drafting; [2,3]Critical analysis and manuscript review; [1,4]Data analysis; Manuscript Editing.

Correspondence:
Muhammad Nasir
Email: drnasir743@gmail.com

Article info:
Received: January 22, 2025
Accepted: March 10, 2025

Cite this article. Ahmad PT, Nasir M, Ali R, Khan I. The Impact of Syrinx on Clinical Outcomes in Arnold-Chiari Malformation: A Retrospective Study. J Islamabad Med Dental Coll. 2025; 14(1): *51-56.*
DOI: https://doi.org/10.35787/jimdc.v14i1.1349

Funding Source: Nil
Conflict of interest: Nil

Introduction

The interplay between syringomyelia and Arnold-Chiari malformation (ACM) has long intrigued neurologists and neurosurgeons due to their potential to cause progressive neurological impairment.[1] ACM, particularly Type I (ACM-I), involves the descent of the cerebellar tonsils below the foramen magnum, disrupting cerebrospinal fluid (CSF) flow and often leading to syrinx formation in the spinal cord. This condition can result in a range of clinical manifestations, from sensory deficits to severe motor dysfunction and scoliosis, which occurs in up to 70% of cases with syringomyelia. Recent advancements in imaging and surgical techniques have improved management, but many questions remain, particularly regarding surgical outcomes and patient prognosis. This study aims to address these gaps by focusing on the incidence and

patterns of syrinx and ACM in a local Pakistani context.[2,3]

ACMs, which can be congenital or acquired, lead to crowding of the cerebellum and brainstem in the posterior cranial fossa, often associated with syringomyelia a fluid-filled cavity within the spinal cord. The incidence of ACM ranges globally from 0.1% to 0.5%, although these numbers are likely underreported due to asymptomatic cases or misdiagnosis.[4] The formation of a syrinx is believed to be due to altered CSF dynamics at the craniovertebral junction, leading to fluid accumulation and spinal cord damage. Advanced imaging techniques, such as phase-contrast MRI, have demonstrated abnormal CSF flow patterns, particularly in patients with syringomyelia.[5]

The clinical presentation of ACM-I with syrinx varies widely, with some patients remaining asymptomatic and others developing severe symptoms, including headaches, neck pain, motor weakness, and sensory loss. Early diagnosis and intervention are crucial to prevent irreversible damage.[6,7] A review of Chiari malformations highlights the underreporting of cases, especially in regions with limited access to advanced diagnostics, which complicates treatment. Posterior fossa decompression has been shown to improve CSF dynamics in patients with ACM-I and syringomyelia, with significant reductions in syrinx size and symptomatic improvement, particularly when performed early.[8,9]

Studies from Pakistan have noted challenges, such as delayed diagnosis and limited follow-up care, due to restricted access to imaging and specialized neurosurgical expertise. These issues are further complicated by regional healthcare disparities, which influence surgical outcomes. For instance, delayed diagnosis often leads to severe complications, such as scoliosis or respiratory difficulties. The pathophysiology of syrinx formation in ACM involves disrupted CSF flow and impaired venous drainage, which exacerbates the pressure gradient across the spinal cord.[4]

Despite advancements in understanding the condition and the role of surgery in restoring CSF flow, significant gaps remain, particularly in regions like Pakistan. The lack of localized data limits the ability to tailor diagnostic and therapeutic strategies to the specific needs of the population. This study seeks to fill these gaps by investigating the incidence, morphological patterns, and clinical outcomes of ACM and syringomyelia in patients treated at Hayatabad Medical Complex, Peshawar.[10]

Methodology

Study Design and Setting: This retrospective study was conducted at the Department of Neurosurgery, Hayatabad Medical Complex, Peshawar, over a period of one year, from January 2023 to December 2023. The study aimed to analyse the incidence trends, morphological patterns, and outcomes of syringomyelia and Arnold-Chiari malformation in patients treated during this timeframe.

Sample Size: A total of 150 patients were included in the study. The sample size was calculated using the WHO formula for sample size determination, considering the prevalence of Chiari malformation with syringomyelia reported in prior studies. For example, a retrospective study identified syrinx in 40% of Arnold-Chiari malformation cases.[3] Assuming a 95% confidence level and a 5% margin of error, this prevalence guided the calculation of our sample size. The patients were divided into two groups: 75 with syringomyelia and 75 with Arnold-Chiari malformation without syrinx, to allow comparative analysis.

Inclusion and Exclusion Criteria: Patients were included if they were diagnosed with Arnold-Chiari malformation, with or without syringomyelia, confirmed by magnetic resonance imaging (MRI). Inclusion also required patients aged 18 and above who underwent neurosurgical intervention or conservative management during the study period. Patients with incomplete medical records, prior neurosurgical treatment for Chiari malformation, or

concurrent conditions unrelated to Arnold-Chiari malformation or syringomyelia, such as traumatic spinal injuries, were excluded from the study.

Data Collection Procedure: Data were collected retrospectively from patient medical records, radiological reports, and surgical documentation. Key variables included patient demographics, clinical presentation, MRI findings (extent of tonsillar herniation, syrinx morphology, and location), surgical details (if applicable), and postoperative outcomes. Each patient's records were anonymised and assigned a unique study ID to ensure confidentiality. Data were independently verified by two researchers to minimize errors and inconsistencies.

Definitions and Assessment Criteria: Arnold-Chiari malformation was defined as herniation of the cerebellar tonsils ≥5 mm below the foramen magnum, based on MRI findings. Syringomyelia was identified as the presence of a fluid-filled cavity within the spinal cord. Clinical improvement was assessed based on documented symptom relief (headaches, motor weakness, and sensory deficits) and radiological improvement on follow-up imaging. Surgical outcomes were categorized as "successful" if postoperative imaging showed syrinx reduction or tonsillar repositioning and "unsuccessful" if no significant change was observed.

Statistical Analysis: Data were analysed using descriptive and inferential statistics. Continuous variables (e.g., age, syrinx size) were summarized as mean ± standard deviation, while categorical variables (e.g., presence or absence of syrinx) were expressed as percentages. Comparative analysis between groups was conducted using appropriate statistical tests, with a significance level set at $p<0.05p < 0.05$. The analysis was performed using a standard statistical software SPSS version 25.

Ethical approval for the study was obtained from the Ethical and Research Committee of Hayatabad Medical Complex, Peshawar. (IRB#1650 dated 12th December 2022) The study adhered to the principles outlined in the Declaration of Helsinki. All data collection and analysis procedures were conducted in compliance with ethical standards to ensure patient confidentiality and data integrity.

Results

The study included a total of 150 patients, with 75 patients diagnosed with Arnold-Chiari malformation and syringomyelia (Group 1) and 75 patients with Arnold-Chiari malformation without syringomyelia (Group 2). The mean age across all participants was 42.5 ± 15.2years.

Syrinx Group Statistics: The distribution of key variables such as age, tonsillar herniation, and syrinx length is summarized in Table I**,** showing the statistical differences between groups. The presence of syrinx significantly correlated with higher tonsillar herniation values ($p<0.001$).

Surgical Outcomes: A breakdown of surgical outcomes in relation to syrinx presence is presented in **Table II**. The chi-square analysis revealed a significant association between surgical outcomes and syrinx presence ($p<0.05$).

Scatterplot of Age and Tonsillar Herniation: The relationship between patient age and tonsillar herniation, segmented by syrinx presence is shown. This scatterplot highlights that patients with syrinx tend to have more pronounced tonsillar herniation across all age groups.

Syrinx Length by Surgical Outcome: Figure 2 demonstrates the distribution of syrinx length across surgical outcomes for patients with syringomyelia. Patients with "Improved" outcomes exhibited smaller syrinx lengths, indicating a possible predictive relationship between syrinx size and treatment success

Table I. Syrinx Group Statistics (n=150)

Syrinx Present	Mean Age (yrs)	Std Age	Mean Tonsillar Herniation (mm)	Std Tonsillar Herniation (mm)	Mean Syrinx Length (mm)	Std Syrinx Length (mm)
No	40.7	14.2	9.2	2.6	0	0
Yes	45.4	14.9	9.8	2.6	27.5	12.1

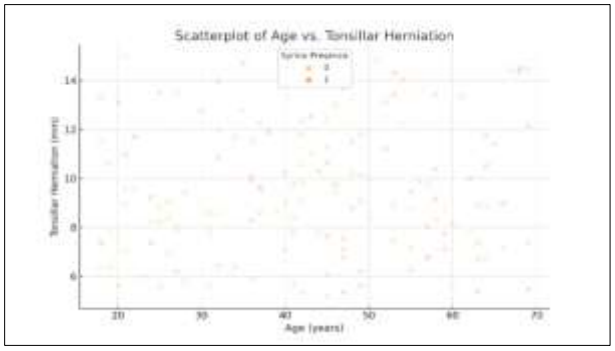

Figure 1. Scatterplot of Age vs. Tonsillar Herniation

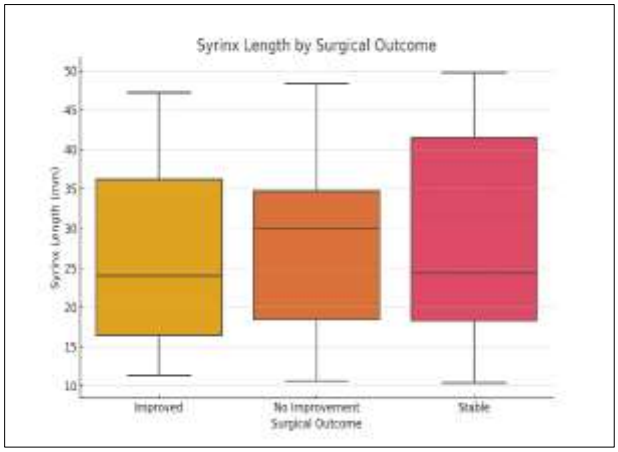

Figure 2. Syrinx Length by Surgical Outcome

Table II. Surgical Outcome Statistics (n=150)				
Surgical Outcome	**Without Syrinx**	**With Syrinx**	**Total**	**P< 0.05**
Improved	0	26	26	
No Improvement	0	20	20	
Not Applicable	81	0	81	
Stable	0	23	23	
All	81	69	150	

Statistical Insights: Statistical analysis revealed significant differences in key parameters between patient groups. Patients with syringomyelia exhibited significantly higher tonsillar herniation values compared to those without syringomyelia ($p<0.001$), indicating a strong correlation between the presence of syrinx and the degree of cerebellar tonsillar descent. Additionally, a chi-square test demonstrated a statistically significant relationship between syrinx presence and surgical outcomes ($p<0.05$), highlighting that the presence of a syrinx may influence the likelihood of achieving favourable post-surgical results. These findings underscore the critical role of syrinx morphology in determining patient prognosis and treatment efficacy.

Discussion

According to the findings of this research, there are notable variations in clinical and surgical characteristics between individuals who have Arnold-Chiari malformation and syringomyelia and those who do not have syringomyelia. Syrinx patients had a larger tonsillar herniation and a more intense degree of symptoms than those without the condition. There was also a substantial association between the existence of syrinx and the results of surgical procedures, which suggests that it plays a role in the efficacy of therapeutic interventions. Specifically with regard to the local population that is being treated at Hayatabad Medical Complex in Peshawar, these results provide essential insights into the etiology and therapy of these illnesses.

Numerous studies have been conducted all over the world to investigate the connection between Arnold-Chiari malformation and syringomyelia. These studies have shed light on the relevance of altered CSF fluid dynamics in the genesis of syrinx via their findings.[4] It was revealed in a research that there is a link between tonsillar descent and syrinx shape in individuals with scoliosis. This further emphasizes the need of early surgical intervention.[3] The results of these studies offer a solid basis for our findings and are consistent with the patterns that have been noticed in our patient cohort.

Comparable trends in surgical outcomes and syrinx resolution have been found in studies conducted in both Europe and North America after posterior fossa decompression. The findings of our investigation are consistent with the findings of a study that verified the prognostic function of tonsillar herniation and syrinx size in determining the effectiveness of surgical procedures.[5,11] In a similar

vein, a research brought to light the fact that the results of surgical procedures might vary greatly depending on certain anatomical and clinical characteristics of the patient.[12]

There are not many research that have been conducted on Arnold-Chiari malformation and syringomyelia in South Asia. This research is among the first to completely analyze the incidence trends and consequences of these illnesses in a Pakistani community. The absence of reliable local data highlights the novelty of this study, which is among the first to do so. Case reports or studies conducted on a smaller scale have been the primary focus of previous research, which has lacked a comprehensive statistical analysis.

Limited access to modern imaging methods and delayed diagnosis have been highlighted by a few studies that have been conducted in South Asia. These research have recorded individual instances or small cohorts.[13,14] The study investigated the effect that posterior fossa decompression has on syringomyelia, however it did not concentrate on the incidence patterns of the disease in larger patient groups or on geographical characteristics.[15]

The Arnold-Chiari malformation has been discussed in local literature on occasion; however, the majority of these works are descriptive rather than analytical in nature. A significant contribution to the existing body of information is made by this research, which incorporates statistical rigor and directly addresses the effect of syrinx on the results of surgical procedures. Syringomyelia and Arnold-Chiari malformation are the subjects of the present research, which focuses on the interaction between the two conditions. This study is directly related to the topic that is being investigated. Findings such as the considerable association between tonsillar herniation and syrinx development not only provide credence to hypotheses that have been established in the worldwide literature, but they also provide novel insights into the background of the area. The substantial correlation between the existence of syrinx and the results of surgical procedures highlights the need of developing individualized care regimens.

Furthermore, the findings indicate that individuals who have syrinx are at a higher risk of experiencing severe clinical symptoms and need therapies that are more specifically focused. The fact that patients with smaller syrinxes had a better percentage of surgical success implies that initiating treatment at an earlier stage might potentially enhance results.[4] The results of this study are in line with those of earlier research that emphasized the need of prompt diagnosis and treatment.

Study Limitations and Future Directions: This study is limited by its retrospective design, which may introduce selection bias and restrict causal inferences. Additionally, the single-centre setting limits the generalizability of findings to other regions in Pakistan or globally. Future research should involve multicentre studies with larger sample sizes to validate these findings. Prospective studies exploring the impact of novel imaging techniques and individualized surgical approaches could further enhance patient care.

Conclusion

This study provides critical insights into the interplay between Arnold-Chiari malformation and syringomyelia, highlighting the significant relationship between syrinx presence and clinical outcomes. Patients with syrinx exhibit higher tonsillar herniation and distinct surgical outcomes compared to those without syringomyelia. These results emphasize the importance of early diagnosis and intervention, which can significantly improve patient outcomes.

Recommendation: Future research should focus on multicentre studies to validate these findings and explore advanced imaging techniques and personalized surgical approaches. Strengthening diagnostic and treatment infrastructure in resource limited settings like Pakistan is essential to ensure equitable healthcare delivery for these conditions.

References

1. Rostorguev E, Kuznetsova NS, Maslov AA, Hatyushin VE, Matevosyan B V., Reznik GA, et al. a rare clinical case of syringomyelia progression in the presence of chiari i malformation following the surgery. South Russ J Cancer 2023;4:44–50.. https://doi.org/10.37748/2686-9039-2023-4-3-5.
2. Sadler B, Kuensting T, Strahle J, Park TS, Smyth M, Limbrick DD, et al. prevalence and impact of underlying diagnosis and comorbidities on chiari 1 malformation. Pediatr Neurol 2020;106:32–7. https://doi.org/10.1016/j.pediatrneurol.2019.12.005
3. Shanmugasundaram S, Viswanathan VK, Shetty AP, Rai N, Hajare S, Kanna RM, et al. Type i arnold chiari malformation with syringomyelia and scoliosis: radiological correlations between tonsillar descent, syrinx morphology and curve characteristics: a retrospective study. Asian Spine J 2023;17:156–65. https://doi.org/10.31616/asj.2021.0483.
4. Capel C, Lantonkpode R, Metanbou S, Peltier J, Balédent O. hemodynamic and hydrodynamic pathophysiology in chiari type 1 malformations: towards understanding the genesis of syrinx. J Clin Med2023;12:5954. https://doi.org/10.3390/jcm12185954
5. Luzzi S, Giotta Lucifero A, Elsawaf Y, Elbabaa SK, Del Maestro M, Savioli G, et al. pulsatile cerebrospinal fluid dynamics in chiari i malformation syringomyelia. J Craniovertebr Junction Spine 2021;12:15–25. https://doi.org/10.4103/jcvjs.JCVJS_42_20.
6. Ravindra VM, Brockmeye DL. Chiari and scoliosis. The Chiari Malformations, Cham: Springer International Publishing; 2020, p. 219–24. https://doi.org/10.1007/978-3-030-44862-2_17.
7. Rodríguez EM. chiari malformations: a review of the current literature. Arch Clin Trials Case Reports 2023. http://.doi.org/10.37191/Mapsci-ACTCR-2(1)-20
8. Mago V, Chakole V, Nisal R, Umate R. a case of anesthetic management of arnold-chiari malformation i: a contest to anesthesiologists. Cureus 2023. http://dx.doi.org/10.7759/cureus.33848.
9. Tosi U, Lara-Reyna J, Chae J, Sepanj R, Souweidane MM, Greenfield JP et al. persistent syringomyelia after posterior fossa decompression for chiari malformation. World Neurosurg 2020;136:454-461.e1. https://doi.org/10.1016/j.wneu.2020.01.148
10. Shukla DA, Gupta DAK, Iyengar DS. result of posterior fossa decompression on syringomyelia in cases of chiary type i malformation. Sch J Appl Med Sci 2020;08:413–6. https://doi.org/10.36347/sjams.2020.v08i02.011.
11. Shahab M. A rare case of arnold chiari malformation evident after a streptococcal throat infection in a young female. J Clin Med Images, Case Reports 2023;3. http://.doi.org/10.7759/cureus.42024.
12. Passias P, Naessig S, Para A, Ahmad W, Pierce K, Janjua Mb, et al. complication rates following chiari malformation surgical management for arnold–chiari type i based on surgical variables: a national perspective. J Craniovertebr Junction Spine 2020;11:169. https://doi.org/10.4103/jcvjs.jcvjs_69_20.
13. Hamal D, Fernandes A, Ghimire P, Wong A. Acute respiratory failure requiring invasive ventilation in adults with congenital syringomyelia/arnold-chiari malformations: a systematic review. Cureus 2024. https://doi.org/10.7759/cureus.70109.
14. Emmert AS, Yahanda AT, Limbrick DD, Mangano FT. 343 Influence of duraplasty on clinical, radiological, and surgical factors associated with syrinx location in children treated for chiari malformation type i and syringomyelia: a park-reeves syringomyelia research consortium study. Neurosurgery 2023;69:54–54. https://doi.org/10.1227/neu.0000000000002375_343.
15. Antkowiak L, Tabakow P. Comparative assessment of three posterior fossa decompression techniques and evaluation of the evidence supporting the efficacy of syrinx shunting and filum terminale sectioning in chiari malformation type i. a systematic review and network meta-analysis. World Neurosurg 2021;152:31–43. https://doi.org/10.1016/j.wneu.2021.05.124.