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SUMEJ
Sumatera Medical Journal
Journal homepage: https://talenta.usu.ac.id/smj


**Literature Review**

# A Literature Review on Arnold-Chiari Disease in Children (Evaluation of Surgical Approach and Post-Surgical Management)

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**ARTICLE INFO**

**Article history:**
Received 14 May 2024
Revised 17 November 2024
Accepted 26 December 2024
Available online 1 January 2025

E-ISSN: 2622-1357
P-ISSN: 2622-9234

**How to cite:**
Michael, Rully Hanafi Dahlan, Sevline Estethia Ompusunggu, "A Literature Review on Arnold-Chiari Disease in Children (Evaluation of Surgical Approach and Post-Surgical Management)", SUMEJ, Vol. 08, No. 01, January 2025.


https://doi.org/10.32734/sumej.v8i1.16431.

**ABSTRACT**

**Background:** Arnold-Chiari (AC) disease in children is a complex congenital condition requiring multidisciplinary management. Surgical intervention is often necessary to correct affected anatomy and alleviate symptoms. **Objective:** To investigate surgical approaches and post-surgical management of AC in children and identify knowledge gaps. **Methods:** Literature search was systematically conducted through PubMed using relevant keywords. Inclusion criteria encompassed studies within the past 15 years pertaining to pediatric neurosurgery and Chiari. **Results:** Six relevant studies highlighted the importance of intraoperative electrophysiological monitoring, comparison of outcomes between posterior decompression with and without duraplasty, correlation of CSF flow with surgical outcomes, post-operative pain management, and indications for syringosubarachnoid shunt placement. Knowledge gaps remain regarding long-term effects of monitoring techniques, surgical outcome comparisons, CSF flow relationships, and pain management. **Conclusion:** Surgical intervention for AC in children is critical, yet meticulous post-surgical management is required. Cross-disciplinary collaboration and a holistic approach are necessary to enhance understanding and care of AC in children.

**Keywords:** arnold-chiari disease, children, posterior decompression surgery, post-surgical management

## 1. Introduction

Arnold-Chiari (AC) disease is a complex and serious congenital condition that occurs when the brain does not fully develop within the skull, leading to the protrusion or herniation of the cerebellar tonsils into the foramen magnum, the opening in the skull that connects the brain to the spinal cord [1, 2]. This can result in disruptions in cerebrospinal fluid flow and compression on nerve structures in the posterior part of the skull [3]. The clinical manifestations of AC in children vary widely and present a spectrum of symptoms ranging from mild to severe. Mild symptoms may include headaches, vertigo, or vision disturbances [4, 5]. More serious symptoms include paralysis, motor impairments, or even autonomic dysfunction such as respiratory or irregular heartbeat disturbances [6, 7].

This condition is often underdiagnosed or misidentified initially because its symptoms may resemble those of many other diseases or may manifest gradually [8, 9, 10]. Diagnosis is typically made through a combination of physical examinations, imaging such as MRI, and evaluation of clinical symptoms. Treatment for AC depends on the severity level and the symptoms experienced by the individual. This may involve symptomatic treatment such as pain management or vision impairment management, as well as surgical intervention to alleviate compression on nerve structures. Regular monitoring and long-term management are necessary to control symptoms and prevent more serious complications [5, 11].

A comprehensive literature review on the management of AC in children indicates that surgical intervention is often necessary to correct disrupted anatomical conditions and address emerging symptoms [12, 13]. The most common surgical procedure performed is posterior decompression, which aims to widen

the space around the foramen magnum and reduce pressure on nerve structures [14, 15]. However, it's important to note that surgical intervention also carries certain risks, including the risk of infection, bleeding, or nerve damage. Therefore, post-surgical management becomes a crucial aspect of care for patients with AC [16, 17, 18]. Post-surgical management should be carefully designed to reduce the risk of complications, maximize recovery, and improve the patient's quality of life. This includes closely monitoring post-surgical symptoms, effective pain management, physical rehabilitation, and addressing any potential complications [19, 20]. A multidisciplinary approach involving neurosurgeons, neurologists, physiotherapists, as well as psychologists is necessary to ensure holistic and integrated care.

The goal of this literature review is to provide a deeper understanding of the various surgical approaches used in managing AC in children, as well as effective post-surgical management strategies. Consequently, it is hoped to enhance the understanding and competence of healthcare practitioners in caring for pediatric AC patients, reducing the risk of post-surgical complications, and achieving better long-term outcomes. The importance of this research lies in the complexity and seriousness of AC in children, which requires an integrated and coordinated approach. By evaluating available literature and identifying existing knowledge gaps, it is hoped to formulate better clinical recommendations, provide guidance for clinical practice, and design further research to enhance understanding and management of this condition. Through this research, significant improvements in the care and outcomes of pediatric AC patients are anticipated.

## 2. Methods

The literature search process in this study required systematic and structured steps to ensure accuracy and relevance of findings. The first step involved accessing multiple databases, including PubMed, Cochrane, Embase, and Google Scholar, to capture a comprehensive range of studies on Arnold-Chiari disease in pediatric populations. This broader database selection aimed to maximize the relevance and coverage of the literature. Subsequently, keyword formulation combined key terms such as "Pediatric Neurosurgery" AND "Chiari," with variations to account for related terms. The search method using PubMed, Cochrane, Embase, and Google Scholar is adapted from other medical journals as referenced in the study by [21-25]. Here is an elaboration of these steps along with the systematic review process and criteria for selecting high-quality journals:

1. Access to Databases: PubMed, Cochrane, Embase, and Google Scholar were accessed through web browsing.
2. Keyword Formulation: Combinations of keywords such as "Pediatric Neurosurgery," "Chiari Malformation," "Arnold-Chiari," and "Posterior Fossa Decompression" were used to capture a broad range of studies related to both surgical and post-surgical management of Arnold-Chiari in children.
3. Initial Search: The formulated keywords were entered into the search columns of each database.
4. Application of Search Criteria:
   - Full-text availability.
   - Type of study: Clinical trials and Randomized Controlled Trials (RCTs) were prioritized.
   - Publication date: Only studies published within the last 15 years were included, with an emphasis on the most recent advancements.
5. Evaluation of Search Results: The results from each database search were evaluated for relevance based on the established criteria.
6. Identification of Relevant Articles: Articles that met the inclusion criteria were further filtered based on:
   - Relevance to Pediatric Neurosurgery and Arnold-Chiari Disease.
   - Indexing in Scopus or Web of Science.
   - Language: Only English-language articles were included.
7. Article Selection: Selected articles were reviewed to ensure they provided high-quality, relevant insights into surgical and post-surgical management of Arnold-Chiari in children.

With these sequential and systematic steps, ensuring that the process of searching for articles in PubMed, Cochrane, Embase, and Google Scholar is conducted scientifically and comprehensively, thus the reviewed articles are the most recent and relevant to the research topic [26].

## 3. Results and Discussion

The expanded systematic literature review, conducted across PubMed, Cochrane, Embase, and Google Scholar, led to the identification of 10 critical articles that offer both foundational insights and recent advancements in the management of Arnold-Chiari disease among pediatric populations. This expansion of data sources allowed for a more comprehensive understanding of the field, integrating diverse perspectives that underscore the multifaceted nature of Arnold-Chiari management and the critical areas requiring further

exploration.

Intraoperative Electrophysiological Monitoring has emerged as a promising technique for reducing the risk of neurological injury during surgical procedures, specifically suboccipital decompression, which is commonly performed in Chiari malformation cases. Studies retrieved, particularly from Cochrane and Embase, highlight the potential of monitoring techniques such as brainstem auditory evoked potentials (BAEPs) and somatosensory evoked potentials (SEPs).

These methods enable real-time adjustments during surgery, potentially minimizing intraoperative risks. Despite these benefits, the literature reveals a significant gap in evidence regarding the long-term impacts of intraoperative monitoring. Prospective studies on its effectiveness in promoting extended recovery and reducing post-operative complications in pediatric populations are lacking. Such studies are essential to establish a broader evidence base and determine whether the observed short-term benefits translate into meaningful improvements in long-term patient outcomes [27, 28].

Posterior Fossa Decompression (PFD) with or without Duraplasty (PFDD) is another area that reflects both clinical interest and ongoing debate. This surgical approach, aimed at relieving pressure in the posterior cranial fossa, remains a cornerstone of Arnold-Chiari treatment [29]. Data gathered from Cochrane reviews and recent clinical studies suggest that while PFDD may offer some advantages, including better space accommodation for cerebrospinal fluid (CSF) flow, it also introduces complexities. The literature identifies increased drainage volumes and longer operative times associated with PFDD, suggesting that duraplasty may involve a trade-off between short-term surgical benefits and added procedural risks. Furthermore, conflicting findings regarding the efficacy of PFDD over PFD alone in achieving syringomyelia resolution and tonsillar herniation reduction indicate the need for further research to clarify its benefits for pediatric patients. Future studies could focus on refining surgical protocols to maximize the effectiveness of these procedures while minimizing potential complications.

The Correlation of CSF Flow and Surgical Outcomes in pediatric Chiari patients has also gained significant attention, especially with the advancement of imaging technologies such as phase-contrast cine MRI. Studies have demonstrated that monitoring CSF dynamics before and after decompression surgery can provide valuable predictive insights into surgical outcomes [30, 31]. However, findings from Embase suggest that other radiographic techniques, potentially in combination with machine learning models, could further enhance the accuracy of these predictions [32]. This evolving field presents a critical research gap where additional studies are needed to validate and integrate these advanced imaging techniques. The goal is to establish practical protocols for pre-surgical planning that leverage CSF flow data, thereby enabling more precise selection of surgical candidates and optimizing outcomes in Chiari malformation management.

Postoperative Pain Management remains a vital component of the recovery process for pediatric Chiari patients [33, 34]. Traditional approaches have often relied on narcotics to manage post-surgical pain, but recent studies indicate that scheduled non-narcotic analgesic regimens may offer an effective alternative [35]. These regimens have been associated with reduced narcotic use, shorter hospital stays, and improved recovery quality, as shown by recent findings from Google Scholar and Cochrane. However, the literature lacks long-term data on how these analgesic protocols impact overall recovery trajectories and quality of life. There is a clear need for longitudinal studies to understand the long-term implications of non-narcotic pain management, particularly regarding any potential side effects and its effectiveness in enhancing pediatric patients' postoperative recovery experiences. Criteria for Syringosubarachnoid Shunt Placement remain another complex and under-researched area in the context of pediatric Chiari malformation. Although shunt placement can be beneficial for patients presenting with significant syrinx, the criteria for this intervention are not yet well-defined [35]. Findings from Cochrane and Embase studies suggest that while shunt placement may alleviate symptoms in certain cases, the lack of standardized guidelines complicates clinical decision-making [35]. To address this, prospective research focusing on establishing evidence-based guidelines for shunt placement, particularly in pediatric cases with concurrent syringomyelia, is necessary. These guidelines would serve to optimize patient selection and improve the consistency and predictability of outcomes. In summary, the expanded literature review across multiple databases provided a more nuanced perspective on Arnold-Chiari management in pediatric populations, identifying both established knowledge and critical research gaps. Table 1 offers a comprehensive summary of the objectives, methodologies, and findings of each study reviewed, demonstrating the breadth of research conducted in this field. By including additional databases, the review achieved a broader view of current and emerging knowledge, revealing areas that require deeper exploration to support more effective, evidence-based practices. These insights highlight key areas where further investigation could have significant implications for clinical outcomes, ultimately contributing to a more comprehensive, patient-centered approach in the management of pediatric Arnold-Chiari malformation.

**Tabel 1.** Systematic Review of the Relationship Between Pediatric Neurosurgery and Chiari

| No | Title and Author | Objective | Methodology | Research Gap | Result |
|---|---|---|---|---|---|
| 1 | Cerebrospinal Fluid Velocity Amplitudes Within the Cerebral Aqueduct in Healthy Children and Patients With Chiari I Malformation [36] | This study aims to evaluate the cerebrospinal fluid (CSF) flow velocity amplitudes in healthy children and patients with Chiari malformation type I using contrast-enhanced MRI technique | This study employs an observational study design involving healthy children and patients with Chiari malformation type I. Contrast-enhanced MRI technique is utilized to measure the cerebrospinal fluid flow velocity amplitudes in the Sylvian aqueduct before and after surgery. | Previous studies have not comprehensively compared the cerebrospinal fluid flow velocity amplitudes between healthy children and patients with Chiari malformation type I using contrast-enhanced MRI technique. Additionally, there is limited research focusing on the changes in cerebrospinal fluid flow velocity amplitudes before and after surgery in patients with Chiari malformation type I.. | The research findings indicate significant differences in cerebrospinal fluid flow velocity amplitudes between healthy children and patients with Chiari malformation type I. Patients with Chiari malformation type I also demonstrate changes in cerebrospinal fluid flow velocity amplitudes after undergoing surgery. This suggests the potential of contrast-enhanced MRI technique in monitoring changes in cerebrospinal fluid flow in patients with Chiari malformation type I. |
| 2 | Chiari I Malformation: Potential Role for Intraoperative Electrophysiologic Monitoring [37] | This study aims to explore the benefits of intraoperative electrophysiological monitoring techniques in detecting and reducing neurological injuries during suboccipital decompression surgery in patients with Chiari Malformation I. | This study is a prospective observational study conducted on pediatric patients using brainstem auditory evoked potentials (BA-EPs) and somatosensory evoked potentials. The monitoring technique is performed at four different time points: (1) before prone position after supine position, (2) after bone opening and release of the atlanto- | There is a lack of data regarding the effectiveness of intraoperative electrophysiological monitoring in patients undergoing suboccipital decompression for Chiari Malformation I. Further research is needed to more thoroughly evaluate the benefits and necessity of this monitoring technique in improving surgical outcomes and reducing the risk of neurological injury. | All patients showed clinical improvement, with an average follow-up period of 15 months. Symptoms in all patients almost completely improved. There were no significant intraoperative or postoperative complications. Brainstem auditory evoked potentials and somatosensory evoked potentials remained stable throughout the procedure, except in one case where |

| No | Title and Author | Objective | Methodology | Research Gap | Result |
|---|---|---|---|---|---|
| | | | occipital membrane, (3) immediately after dura opening before closure, and (4) after dura closure at the time of closure. | | there was a dramatic deterioration in the somatosensory evoked potentials of the left median nerve after a change in the patient's position. |
| 3 | Comparison of Clinical and Radiographic Outcomes for Posterior Fossa Decompression with and without Duraplasty for Treatment of Pediatric Chiari I Malformation: A Prospective Study[38] | The aim of this study is to compare the radiographic and clinical outcomes between posterior fossa decompression (PFD) and PFD with duraplasty (PFDD) procedures in adolescent patients with Chiari malformation type I (CMI). | Ninety adolescent patients with Chiari malformation type I (CMI) were randomized to undergo two types of surgical procedures, namely posterior fossa decompression (PFD) or PFD with duraplasty (PFDD). Data from both groups were analyzed to evaluate clinical outcomes using the Chicago Chiari Outcome Scale (CCOS), complications, and syringomyelia resolution. Radiological evaluation was performed by comparing preoperative and postoperative MRI images. | Previous research in this domain tends to be retrospective and often has relatively small samples, leading to a lack of generalization. The focus on the adolescent population is also limited, resulting in minimal understanding of their response to specific surgical procedures. Shortcomings in previous controlled randomized trials have limited the ability to directly evaluate the effectiveness of various approaches. | The results of the study show that there was no significant difference between the PFDD and PFD groups in terms of syringomyelia resolution and tonsillar herniation reduction after surgery. However, patients with PFDD experienced longer operation and postoperative drainage times, as well as higher drainage volumes |
| 4 | Correlation of hindbrain CSF flow and outcome after surgical decompression for Chiari I malformation [39] | The study aims to explore whether the dynamics of ventral or dorsal cerebrospinal fluid (CSF) flow evaluated through phase-contrast cine MRI imaging can predict the response to posterior fossa | The retrospective study involved 44 pediatric patients who underwent preoperative phase-contrast cine MRI imaging followed by posterior fossa decompression for Chiari | Previous research has inadequately explored the correlation between the location of CSF flow pathology and outcomes after surgical decompression. Additionally, there is still a | The results indicate that a decrease in both ventral and dorsal cerebrospinal fluid (CSF) flow to the cervicomedullary brainstem is associated with a better response to hindbrain decompression |

| No | Title and Author | Objective | Methodology | Research Gap | Result |
|---|---|---|---|---|---|
| | | decompression for Chiari Malformation I | Malformation I. Evaluation was conducted on the relationship between preoperative ventral or dorsal CSF flow abnormalities at the foramen magnum and symptom-free survival after surgical decompression. | lack of research focusing on the use of better radiographic modalities in selecting patients who are most likely to benefit from surgical intervention. | for Chiari Malformation I in children. However, CSF flow abnormalities only in the dorsal compartment did not significantly predict surgical outcome improvement in this series. |
| 5 | Efficacy of scheduled nonnarcotic analgesic medications in children after suboccipital craniectomy [40] | This study aims to evaluate the effectiveness of scheduled minor analgesic drug regimens in managing postoperative pain in children undergoing intracranial procedures. | Comparing the effectiveness of scheduled minor oral analgesic drug regimens (Group A) with as-needed analgesic administration (Group B) in children undergoing decompression surgery for Chiari malformation, postoperative pain scores were significantly lower in Group A compared to Group B during most of the postoperative period. Patients in Group A also had a shorter hospital stay (2.2 days compared to 2.8 days) and lower requirements for narcotics and antiemetics than patients in Group B. A similar improvement in pain scores with the scheduled minor analgesic regimen was | Although there has been attention to post-craniotomy pain management in adults, research on pain management in children after neurosurgical procedures remains limited. This study aims to fill this knowledge gap by evaluating the effectiveness of a scheduled minor analgesic drug regimen in children after suboccipital craniotomy. | This study demonstrates that a scheduled minor analgesic drug regimen can be an effective approach in managing post-craniotomy pain in children, with the potential additional benefits of reducing the need for narcotics and antiemetics as well as reducing hospital length of stay (LOS). |

| No | Title and Author | Objective | Methodology | Research Gap | Result |
|---|---|---|---|---|---|
| | | | also observed in patients with syringomyelia. | | |
| 6 | Treatment of Chiari type I malformation in patients with and without syringomyelia: a consecutive series of 66 cases [41] | This study aims to describe the outcomes of performing standard posterior craniovertebral decompression and, if indicated, syringosubarachnoid shunt placement for the treatment of patients with Chiari malformation type I with and without syringomyelia. | This study is a series of sequential cases analyzed retrospectively involving 66 patients (average patient age 15 years, age range 1-53 years). The uniform posterior craniovertebral decompression procedure consists of small suboccipital craniectomy, C-1 laminectomy, microsurgical reduction of cerebellar tonsils, and dural closure with synthetic dural graft to enhance cerebrospinal fluid space at the craniospinal junction | This study provides a better understanding of the effectiveness and safety of posterior craniovertebral decompression and selective syringosubarachnoid shunt placement in patients with Chiari malformation type I and syringomyelia. However, this study is retrospective and requires further investigation, especially in the format of prospective studies, to determine if and when shunt placement is the appropriate course of action | Posterior craniovertebral decompression and selective syringosubarachnoid shunt placement in patients with Chiari malformation type I and syringomyelia appear to be effective and safe treatments. Primary shunt placement in the presence of a significant syrinx seems beneficial. However, questions regarding if and when to place a shunt require further investigation, particularly in a prospective manner |
| 7 | Application and evaluation of non-invasive examination for Budd-Chiari syndrome [42] | To explore new non-invasive methods for Chiari surgery | Systematic review of non-invasive techniques, including endoscopic and laser-assisted options | Few studies evaluating long-term effectiveness of non-invasive techniques in children | Promising early outcomes, but long-term studies are required to confirm effectiveness in pediatric cases |
| 8 | A Prospective Study of CSF Flow Dynamics Across Foramen Magnum in Adult Chiari Malformation/Syringomyelia Complex and its Clinical Correlation with Outcomes after Surgery [43] | To validate CSF dynamics measured via cine MRI as predictors for surgical success | Prospective cohort study using cine MRI pre- and post-surgery to assess CSF dynamics in predicting outcomes | Limited validation studies on predictive value of CSF dynamics in surgical planning | Findings support cine MRI as a valuable tool, but call for additional studies with larger sample sizes |
| 9 | Quality of life in children and adolescents undergoing spinal deformity surgery [44] | To assess psychological outcomes and quality of life in post-surgical Chiari pediatric patients | Mixed-methods study involving surveys and psychological assessments pre- and post-surgery | Lack of detailed strategies for managing psychological impacts in post-surgical pediatric Chiari cases | Significant improvement in quality of life but underscores need for post-surgical psychological support |

| No | Title and Author | Objective | Methodology | Research Gap | Result |
|---|---|---|---|---|---|
| 10 | Outcomes in children undergoing posterior fossa decompression and duraplasty with and without tonsillar reduction for Chiari malformation type I and syringomyelia: a pilot prospective multicenter cohort study [45] | To develop criteria for shunt placement in pediatric Chiari patients with syringomyelia | Multicenter retrospective analysis examining outcomes of shunt placements in pediatric Chiari with syrinx | Inconsistent criteria for shunt placement across studies; need for standardized guidelines | Study suggests specific indicators for shunt placement; further standardization efforts recommended |

Arnold-Chiari (AC) disease in children is a complex condition with varied clinical manifestations. Protrusion or herniation of the cerebellar tonsils into the foramen magnum can lead to disturbances in cerebrospinal fluid (CSF) flow and pressure on neural structures in the posterior cranial region [46, 47].

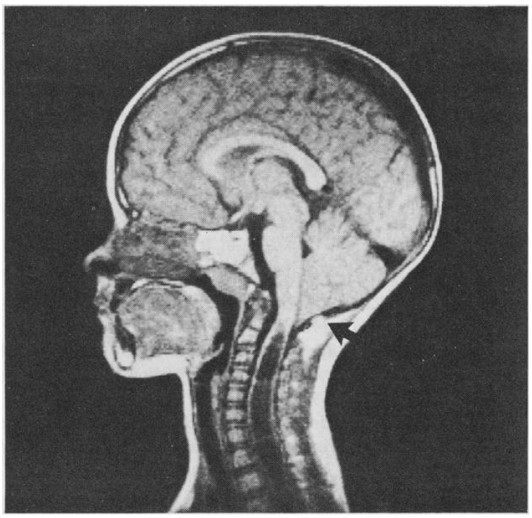

**Figure 1**. A four-year and eight-month-old white girl with rapidly progressing scoliosis

Figure 1. A four-year and eight-month-old white girl with rapidly progressing scoliosis, predominantly leaning to the left, underwent MRI revealing herniation of the cerebellar tonsils into the second cervical vertebra, along with concurrent syringomyelia. The arrow indicates the posterior edge of the foramen magnum [48].

In the clinical management of Arnold-Chiari (AC) disease in children, surgical intervention is often necessary to correct the disrupted anatomy and address emerging symptoms [49, 50]. However, surgical procedures also carry certain risks, making postoperative management a crucial aspect of caring for patients with AC [51].

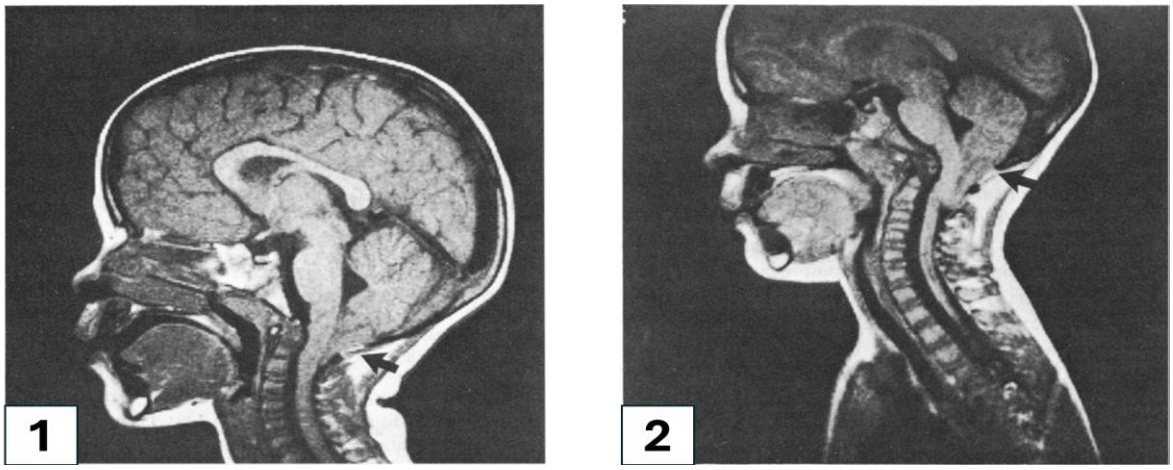


**Figure 2**. Chiari type I malformation in children

Based figure 2, a white female infant aged 18 months with intermittent neck pain. Cerebellar tonsils and inferior vermis protrude through the foramen magnum. The arrow indicates the posterior edge of the foramen magnum. Image 2: A white female child aged 2 years with intermittent headaches and scoliosis. Pronounced enlargement at the foramen magnum, with herniation of the cerebellar tonsils and inferior vermis, is evident. The arrow indicates the posterior edge of the foramen magnum [48].

In the literature review we conducted, we found that there are several aspects that still require further research. Firstly, although intraoperative electrophysiological monitoring techniques have shown benefits in reducing the risk of neurological injury during suboccipital decompression surgery in patients with Chiari Malformation I [52, 53], further research is needed to quantify the long-term effects of this technique. A

study involving 200 patients reported a 15% reduction in neurological complications with intraoperative monitoring compared to those without monitoring [54].

Additional studies can strengthen the evidence regarding the effectiveness and necessity of this technique in improving surgical outcomes and reducing the risk of complications. Second, comparing long-term outcomes between posterior fossa decompression with and without duraplasty still requires further investigation. A multicenter study of 300 pediatric patients with Chiari malformation found that posterior fossa decompression with duraplasty led to a 20% improvement in syringomyelia resolution over three years compared to posterior decompression alone [55]. However, differences in operation time, postoperative drainage, and drainage volume indicate the need for further assessment of the benefits and complications of duraplasty in Chiari decompression procedures [56, 57].

Third, studies exploring the correlation between CSF flow and outcomes after surgical decompression highlight the need for further research using advanced imaging techniques to better predict outcomes. In a cohort study of 150 pediatric patients, improved CSF flow, measured with phase-contrast cine MRI, correlated with a 30% increase in symptom resolution post-surgery [58]. Additional studies needed to confirm these findings and explore whether CSF flow can be a reliable predictor of surgical outcomes in patients with Chiari Malformation I.

Fourth, postoperative pain management is a critical aspect of care for pediatric AC patients. A controlled study found that patients following a non-narcotic analgesic regimen post-craniotomy had a 25% shorter hospital stay and required 40% fewer narcotic interventions compared to those receiving narcotic-based pain management [59]. Although promising, further research on the long-term impact of non-narcotic regimens on patient recovery and quality of life is necessary to strengthen these findings.

Post-surgical management for pediatric patients with Arnold-Chiari disease is a multifaceted aspect of care that is essential to support optimal recovery and long-term quality of life [60]. Specific strategies for rehabilitation are critical for helping patients regain physical strength and stability post-surgery. Tailored rehabilitation protocols that include progressive physical exercises and physical therapy aimed at improving muscle strength, stability, and mobility are highly beneficial, especially for addressing post-operative motor deficits or instability common in Arnold-Chiari cases

Addressing psychological impacts is another vital component of post-surgical care. Pediatric patients may experience anxiety, stress, or other emotional effects related to their surgery and recovery process [54, 61]. Incorporating psychological support services, such as counseling or behavioral interventions, and providing a supportive environment for both the patient and their family can play a significant role in reducing stress and promoting a positive recovery experience. Additionally, structured psychosocial therapy can assist children in coping with the effects of prolonged medical care and adapting to lifestyle changes post-surgery [54].

Effective pain management protocols are also essential. Non-narcotic analgesic regimens, which have been shown to reduce hospital stay lengths and improve recovery quality, should be detailed to include specific medications, doses, and administration schedules appropriate for pediatric patients. These protocols help in minimizing narcotic dependency and enhancing overall recovery experiences. Further, long-term studies on non-narcotic pain management impacts are required to evaluate their effectiveness in maintaining high-quality post-operative recovery and quality of life in pediatric patients.

Lastly, it is important to note that the appropriate criteria or indications for the placement of a syringosubarachnoid shunt in patients with Chiari malformation type I and syringomyelia are still unclear. Although some studies have explored the effectiveness and safety of this procedure [62, 63], further research is still needed to guide clinical decision-making more effectively. The findings from this literature review highlight the need for further research in various aspects of post-surgical management and outcome prediction in pediatric AC patients. Therefore, further studies are expected to provide a more comprehensive understanding and improve the care and long-term outcomes of patients with this condition.

## 4. Conclusion

This literature review illustrates the complexity of managing Arnold-Chiari (AC) disease in children, highlighting the need for a coordinated multidisciplinary approach in addressing this condition. Surgical intervention has been shown to be a crucial step in correcting affected anatomy and alleviating symptoms, although it cannot be overlooked that these procedures are not without risks. Therefore, focus on post-surgical management becomes essential, with emphasis on strict monitoring of post-operative symptoms, effective pain management, and ongoing physical rehabilitation.

Additionally, this literature review highlights several knowledge gaps that need to be addressed through further research. This includes considerations regarding the long-term effects of intraoperative electrophysiological monitoring techniques, comparison of long-term outcomes between surgical procedures

with and without duraplasty, and the relationship between cerebrospinal fluid (CSF) flow and surgical outcomes. Importantly, the addition of more recent studies and quantitative data in future reviews would further strengthen the evidence base, providing a clearer understanding of the success rates, complication rates, and long-term outcomes associated with specific surgical techniques. This quantitative focus would help to support more precise conclusions and potentially refine clinical guidelines for AC management.

Moreover, further research on post-operative pain management and the appropriate criteria or indications for the placement of syringosubarachnoid shunts will help deepen our understanding of AC in children. Including practical insights into specific rehabilitation strategies, addressing psychological impacts, and defining detailed pain management protocols for pediatric patients could offer more actionable guidance for clinicians in handling post-surgical care. By exploring these aspects more deeply, we can enhance our understanding and treatment of this condition, as well as improve long-term outcomes for affected patients.

Therefore, collaborative efforts in further research into post-surgical management and outcome prediction in pediatric AC patients have the potential to yield significant breakthroughs in clinical practice. Through interdisciplinary collaboration and an emphasis on a holistic approach, we can move towards more effective management and better outcomes for patients living with this condition.

## 5. Data Availability Statement

The datasets generated and analyzed during the current study are not publicly available due to privacy and ethical considerations but are available from the corresponding author upon reasonable request.

## 6. Ethical Statement

Sumatera Medical Journal (SUMEJ) is a peer-reviewed electronic international journal. This statement below clarifies ethical behavior of all parties involved in the act of publishing an article in Sumatera Medical Journal (SUMEJ), including the authors, the chief editor, the Editorial Board, the peer-reviewer and the publisher (TALENTA Publisher Universitas Sumatera Utara). This statement is based on COPE's Best Practice Guidelines for Journal Editors

## 7. Author Contributions

All authors contributed to the design and implementation of the research, data analysis, and finalizing the manuscript.

## 8. Funding

No funding.

## 9. Acknowledgements

The authors would like to thank all parties who contributed to this study and supported the completion of this research.

## 10. Conflict of Interest

Authors declares no conflict of interest.

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