Advances in the Understanding and Management of Chiari I Malformation: From Structural Anomaly to Multidimensional Disorder

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Received: May 30, 2026 Accepted: June27, 2026 Published: June 30,2026

doi:10.65343/jiss.v2i1.108 URL: https://doi.org/10.65343/jiss.v2i1.108

Abstract

Background: Chiari I malformation (CM-I) has traditionally been defined as a structural abnormality characterized by caudal displacement of the cerebellar tonsils below the foramen magnum. However, increasing use of advanced neuroimaging and the recognition of asymptomatic cases have challenged this purely anatomical definition. Emerging evidence suggests that CM-I represents a heterogeneous disorder involving cerebrospinal fluid (CSF) dynamics, craniovertebral morphology, and genetic factors.

Methods: A systematic review was conducted according to PRISMA 2020. PubMed/MEDLINE, Scopus, and Web of Science were searched for English-language human studies published from January 1, 2000, to March 31, 2026. Studies addressing pathophysiology, imaging, classification, natural history, or management of CM-I were included. Due to heterogeneity in study design, populations, imaging protocols, and outcomes, a qualitative synthesis was performed.

Results: A total of 124 studies were included in the qualitative synthesis. Current evidence demonstrates that symptom severity in CM-I correlates poorly with the degree of tonsillar descent, highlighting the limitations of traditional diagnostic thresholds. Disruption of CSF flow at the craniovertebral junction has emerged as a central mechanism underlying symptom development and syringomyelia formation. AI-driven analyses have been explored as tools to identify potential phenotypic subgroups and may improve prognostic stratification. Longitudinal studies indicate that while many patients remain stable, a subset experiences progressive neurological decline associated with identifiable risk factors. Surgical management, particularly posterior fossa decompression, remains the primary treatment for symptomatic patients, with evolving techniques aimed at reducing morbidity and improving outcomes.

Conclusion: Chiari I malformation is increasingly recognized as a multifactorial and heterogeneous condition rather than a purely structural anomaly. Integration of advanced imaging, computational modeling, and clinical data is essential for accurate diagnosis and individualized management. Future research should focus on refining phenotypic classification, identifying predictive biomarkers, and advancing precision medicine approaches to optimize patient outcomes.

Keywords: Chiari I malformation, cerebrospinal fluid dynamics, syringomyelia, neuroimaging, artificial intelligence, posterior fossa decompression

1. Introduction

Chiari I malformation (CM-I) is traditionally defined as a structural abnormality of the craniovertebral junction characterized by caudal displacement of the cerebellar tonsils through the foramen magnum. Historically, a descent of ≥5 mm has been used as the principal radiological criterion; however, contemporary evidence indicates that this threshold is overly simplistic and does not adequately reflect the clinical and physiological complexity of the disorder (Bhadelia, Chang, Oshinski, & Loth, 2023, August; Abdallah, Çınar, Gündağ, Güler, & Emel, 2023, March). Increasing use of advanced neuroimaging techniques has revealed that CM-I encompasses a broad spectrum of anatomical and functional alterations rather than a uniform entity (Collins, John, Daniel, Garza, Nagy, & Jacob, 2022, December; Nishikawa, Bolognese, Yamagata, Naito, Sakamoto, Hara, et al., 2022, September 15). (See Figure 1).

One of the most important paradigm shifts in recent years is the recognition that symptom severity correlates poorly with the degree of tonsillar descent. Studies incorporating dynamic imaging have demonstrated that cerebrospinal fluid (CSF) flow abnormalities at the craniovertebral junction play a central role in symptom

generation, suggesting that CM-I should be understood as a disorder of CSF dynamics rather than purely a static anatomical defect (Nair, & Rajshekhar, 2022, December; Ravindra, Iyer, Yahanda, Bollo, Zhu, Joyce, et al., 2021). This concept is further supported by morphometric analyses showing that posterior fossa hypoplasia and craniovertebral junction configuration contribute significantly to disease expression (Mantha, Coulthard, & Campbell, 2021; Marianayagam, Chae, Hussain, Cruz, Baaj, Härtl, et al., 2021, December 6).

Advanced imaging modalities, particularly cine phase-contrast MRI, have provided critical insights into altered CSF flow patterns, including bidirectional flow disturbances and pressure dissociation between intracranial and spinal compartments (Chae, Haghdel, Kelly, Cruz, Wood, Luhmann, et al., 2021, November; King, Liu, Russo, Jayasekara, Stoodley, & Di Ieva, 2024, May 22). These abnormalities are strongly associated with the development of syringomyelia, a common and clinically significant complication of CM-I (Loftus, Wassef, & Ellika, 2024; Yadav, Prasad, Sahu, et al., 2024). The pathophysiological link between CSF obstruction and syrinx formation has been reinforced by both imaging and surgical outcome studies (Khafaji, Sippl, Prell, & Linsler, 2025; McGirt, Atiba, Attenello, Wasserman, Datoo, Gathinji, et al., 2008, July).

Beyond CSF dynamics, structural and biomechanical factors such as clivo-axial angle abnormalities, ventral brainstem compression, and craniocervical instability have emerged as key contributors to disease heterogeneity (Prasad, Azmil Farid, Wen, Zhang, & Kaliaperumal, 2025; Thakar, Kanneganti, Talla Nwotchouang, Salem, Eppelheimer, Loth, et al., 2022, January). Quantitative morphometric studies, including volumetric analysis of the posterior fossa and automated imaging techniques, have further refined the understanding of anatomical variability in CM-I (Kobets, Alavi, Ahmad, Castillo, Young, Minuti, et al., 2024; Bogdanov, Faizutdinova, & Heiss, 2026). These findings underscore the importance of integrating structural and functional assessments in the diagnostic evaluation (Houston, Rogers, Kirkpatrick, Robison Throm, Burgess, et al., 2026; Bogdanov, Faizutdinova, & Heiss, 2021, November).

Recent advances have also highlighted the role of emerging technologies in redefining CM-I. Artificial intelligence (AI)-based approaches have been explored as tools to identify potential phenotypic subgroups by integrating imaging, clinical, and demographic data, thereby challenging traditional binary classifications (Nozaleda, Parras-Martos, Yamamoto, Coenen, Martínez-Bazán, Olivares-Granados, et al., 2026). These data-driven models may improve prognostic stratification and may facilitate personalized treatment strategies in the near future (Staartjes, Öhlén, Lilja, et al., 2025).

From a clinical perspective, CM-I exhibits considerable variability in presentation, ranging from asymptomatic incidental findings to severe neurological impairment. Longitudinal studies indicate that while many patients remain stable over time, a subset demonstrates progressive disease associated with identifiable risk factors, including syringomyelia and abnormal CSF flow dynamics (Ventureyra, Aziz, & Vassilyadi, 2003, February; Aggarwal, Jindal, Rohilla, & Hitesh, 2020). This variability has important implications for clinical decision-making and underscores the need for risk-adapted management strategies (Rindler, & Chern, 2020).

Surgical intervention, particularly posterior fossa decompression, remains the cornerstone of treatment for symptomatic patients. Contemporary surgical approaches aim not only to relieve neural compression but also to restore physiological CSF flow, with increasing emphasis on minimizing morbidity through less invasive techniques and intraoperative imaging guidance (Basaran, Bozdogan, Senol, Gundogan, & Isik, 2021, September; Bogdanov, Faizutdinova, & Heiss, 2022, September 17). Advances in surgical planning and outcome prediction continue to evolve alongside improvements in imaging and computational modeling (Wang, Li, Han, Hu, Pang, Li, et al., 2024).

In addition to structural and neurological manifestations, there is growing recognition of broader clinical dimensions of CM-I, including neurocognitive and psychosocial effects. These aspects further support the concept of CM-I as a multidimensional disorder requiring comprehensive evaluation (Lo, Shiino, Shapiro, Hines, Komune, Donofrio, et al., 2026).

Overall, the current body of evidence supports a shift from a purely anatomical definition toward an integrated model that incorporates CSF dynamics, craniovertebral morphology, and emerging computational insights. This evolving framework provides the basis for a more precise and individualized approach to diagnosis and management (Tubbs, Wellons, Blount, Grabb, & Oakes, 2003; Knight, & De Jesus, 2026, January; Alhosani, & Gachechiladze, 2026).

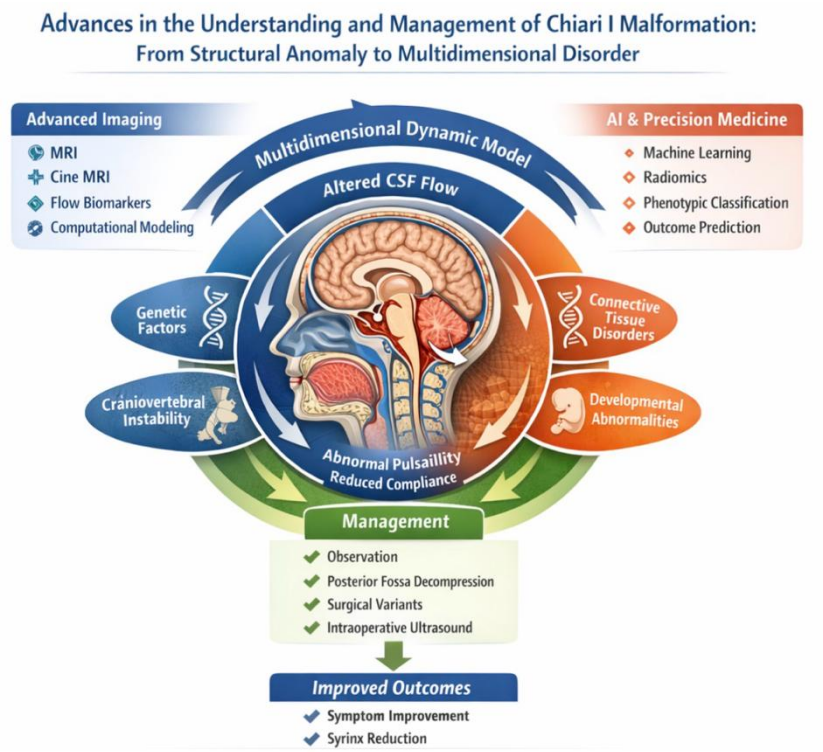


Figure 1. Advances in the Understanding and Management of Chiari I Malformation: From Structural Anomaly to Multidimensional Disorder

Chiari I malformation is illustrated as a multidimensional disorder involving structural abnormalities, altered CSF dynamics, and genetic and developmental factors, integrated with advanced imaging and artificial intelligence to guide management and improve outcomes.

Source: Own elaboration.

2. Methods

2.1 Study Design

This systematic review was conducted in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA 2020) statement, ensuring methodological transparency, reproducibility, and comprehensive reporting of the study selection process and data synthesis. The review protocol was not registered in PROSPERO due to its integrative and multidisciplinary scope, which included heterogeneous study designs and emerging computational methodologies.

2.2 Search Strategy

A structured and comprehensive literature search was performed using three major biomedical databases: PubMed/MEDLINE, Scopus, and Web of Science. The search encompassed studies published between January 1, 2000, and March 31, 2026. The search strategy was designed to maximize sensitivity and specificity by combining controlled vocabulary terms (Medical Subject Headings, MeSH) with free-text keywords, using Boolean operators (AND, OR). The core search string included the following terms: ("Chiari I malformation" OR "cerebellar tonsillar ectopia") AND ("cerebrospinal fluid dynamics" OR "CSF flow" OR "cine MRI") AND ("syringomyelia" OR "posterior fossa decompression" OR "management" OR "artificial intelligence"). Filters were applied to restrict results to human studies published in English. Additionally, manual screening of the reference lists of selected articles was performed to identify potentially relevant studies not captured in the initial database search. The final search was conducted on March 31, 2026.

To ensure reproducibility, the search strategy was defined a priori and applied consistently across databases. Searches were conducted using a combination of controlled vocabulary terms (Medical Subject Headings, MeSH) and free-text keywords. The following fields were queried: MeSH terms and Title/Abstract in PubMed/MEDLINE, TITLE-ABS-KEY in Scopus, and Topic (TS) in Web of Science. Filters applied across all databases included human studies and English language. No restrictions on study design were applied at the search stage.

The core search strategy combined the following conceptual domains: (1) Chiari I malformation and cerebellar tonsillar ectopia; (2) cerebrospinal fluid dynamics, CSF flow, and cine MRI; and (3) clinical outcomes, syringomyelia, management, and artificial intelligence. These domains were combined using Boolean operators (AND, OR).

Given the breadth of the topic, the search was structured into thematic blocks addressing pathophysiology, imaging, clinical presentation, and management, which were combined to maximize sensitivity while maintaining specificity.

The study selection process is illustrated in the PRISMA 2020 flow diagram (Figure 3). The search yielded a total of 1,918 records, of which 1,842 were identified through database searching and 76 through additional sources. After removal of 412 duplicate records, 1,506 unique studies remained for title and abstract screening. At this stage, 1,248 records were excluded due to lack of relevance to Chiari I malformation, absence of original data, or failure to meet predefined inclusion criteria. Subsequently, 258 full-text articles were assessed for eligibility. Of these, 134 studies were excluded for the following reasons: non-relevant outcomes (n = 52), case reports or low-quality evidence (n = 41), studies focused exclusively on pediatric syndromic conditions without clear differentiation of Chiari I malformation (n = 23), and incomplete or insufficient data (n = 18). Ultimately, 124 studies were included in the qualitative synthesis.

The functional alterations in cerebrospinal fluid (CSF) dynamics at the craniovertebral junction, as assessed by cine phase-contrast MRI, are illustrated in Figure 2.

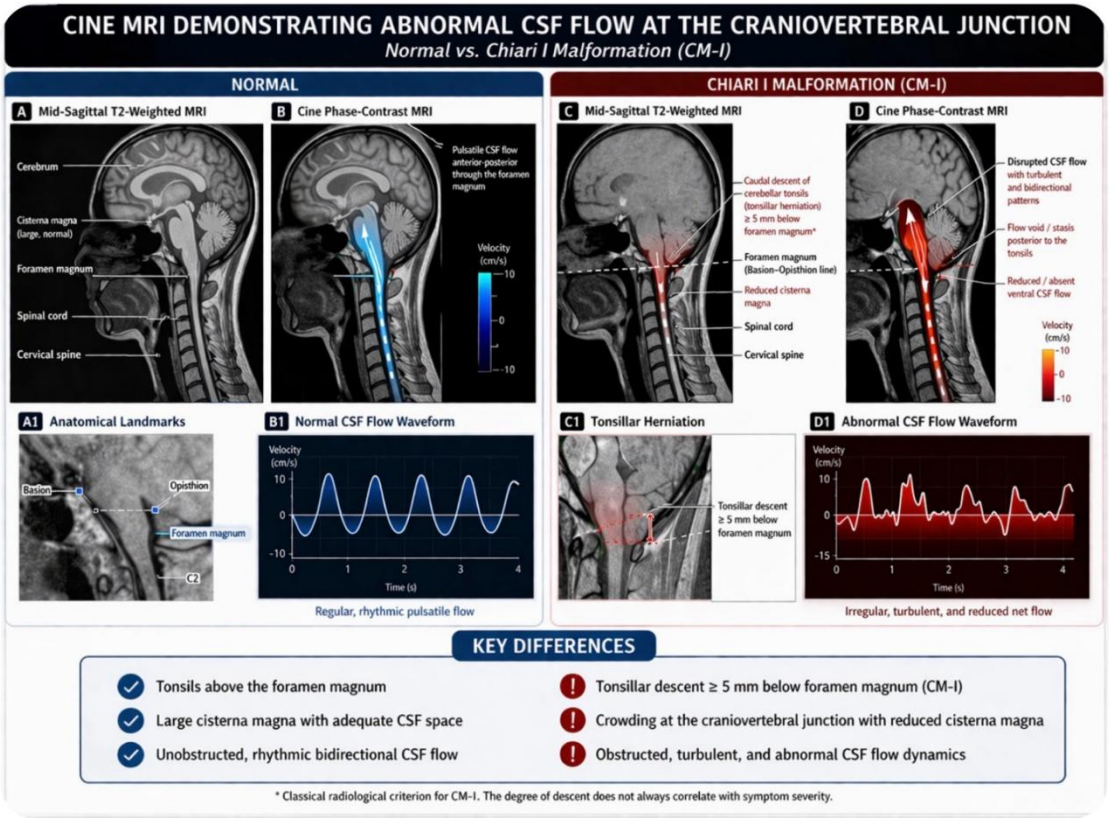


Figure 2. Cine phase-contrast MRI demonstrating cerebrospinal fluid flow dynamics at the craniovertebral junction: comparison between normal physiology and Chiari I malformation

Mid-sagittal T2-weighted and cine phase-contrast magnetic resonance imaging (MRI) illustrating cerebrospinal fluid (CSF) flow at the craniovertebral junction under normal conditions (left panels) and in Chiari I malformation (CM-I) (right panels). In the normal state, cerebellar tonsils are positioned at or above the level of the foramen magnum (basion–opisthion line), with preservation of the cisterna magna and unobstructed, symmetric, bidirectional CSF flow across the foramen magnum, characterized by smooth, rhythmic pulsatility on velocity waveforms. In CM-I, there is caudal descent of the cerebellar tonsils ≥5 mm below the foramen magnum, associated with posterior fossa crowding and reduction of the cisterna magna. These structural changes result in obstruction of CSF pathways, leading to abnormal flow patterns characterized by turbulence, flow jets,

reduced ventral flow, and areas of flow void or stasis. Cine MRI waveforms demonstrate irregular, dampened, and asymmetric pulsatility, reflecting altered craniospinal compliance and pressure dissociation between intracranial and spinal compartments. These findings highlight the central role of CSF hydrodynamics in the pathophysiology of CM-I and support the concept of the disorder as a dynamic, functional condition rather than a purely anatomical abnormality.

Source: Own elaboration.

2.3 Eligibility Criteria

Eligibility criteria were defined a priori. Studies were included if they were original research articles, including prospective or retrospective cohort studies, case-control studies, or clinical trials, involving patients with radiologically or clinically confirmed Chiari I malformation. Chiari I malformation was defined according to radiological criteria as cerebellar tonsillar descent ≥5 mm below the foramen magnum, although studies using alternative or functional definitions were also considered. Additionally, studies were required to report data on at least one of the following domains: pathophysiology, imaging, classification, natural history, or management. Exclusion criteria comprised case reports and small case series with fewer than 10 patients, studies focusing exclusively on secondary or syndromic Chiari malformations, review articles, editorials, conference abstracts without primary data, and studies lacking extractable or clinically relevant information.

2.4 Study Selection

Study selection was performed by a single reviewer (the author), following a two-stage screening process consisting of title and abstract review, followed by full-text assessment. Given the single-reviewer design, strict adherence to predefined inclusion and exclusion criteria was maintained throughout the process to minimize potential selection bias and ensure methodological consistency. To mitigate potential selection bias inherent to a single-reviewer design, predefined eligibility criteria were strictly applied, and the screening process was conducted systematically and consistently across all records.

2.5 Data Extraction and Synthesis

Data extraction was conducted systematically using a standardized data collection form developed specifically for this review. Extracted variables included study characteristics (first author, year of publication, country, and study design), population data (sample size, age group categorized as pediatric, adult, or mixed, and sex distribution when available), and diagnostic and imaging variables (degree of tonsillar descent in millimeters, presence of syringomyelia, imaging modality including MRI and cine MRI, and CSF flow characteristics). Additional pathophysiological variables included evidence of CSF flow obstruction, craniovertebral junction abnormalities, and associated structural findings. Management-related data encompassed conservative versus surgical treatment approaches and specific surgical techniques employed. Outcome measures included clinical improvement, radiological outcomes such as syrinx reduction, complications, and duration of follow-up. When data were incomplete or inconsistently reported, studies were included only if sufficient information could be extracted for qualitative interpretation. Due to the large number of included studies (n = 124), representative studies are presented in summary tables, while the full dataset supporting the findings of this study is available from the corresponding author upon reasonable request.

2.6 Risk of Bias Assessment

The methodological quality of included studies was assessed using the Newcastle–Ottawa Scale (NOS) for observational studies. Only eligible cohort and case-control studies included in the qualitative synthesis were evaluated using this tool. The NOS domains assessed included selection, comparability, and outcome or exposure. Systematic reviews, narrative reviews, and conceptual or methodological articles were not included in the formal risk-of-bias assessment and were used only for contextual interpretation.

2.7 Quality of Evidence

The assessment was conducted by a single reviewer using predefined criteria. In cases of uncertainty, full-text reassessment was performed and conservative judgments were applied to ensure consistency. No studies were excluded based on quality scores; however, methodological quality was considered during interpretation of the findings.

Due to substantial heterogeneity in study designs, patient populations, imaging methodologies, and outcome measures, a quantitative meta-analysis was not performed. Heterogeneity was observed in study design, imaging protocols, patient populations, and outcome definitions, precluding meaningful quantitative synthesis. Instead, a structured qualitative synthesis was undertaken. Findings were organized into key thematic domains, including cerebrospinal fluid dynamics, advances in imaging, phenotypic classification, natural history, and management strategies. These domains were selected to capture the multidimensional nature of CM-I, including structural,

functional, and therapeutic aspects. This integrative approach enabled the synthesis of structural, functional, and emerging computational evidence, providing a comprehensive and clinically meaningful interpretation of the current state of knowledge on Chiari I malformation.

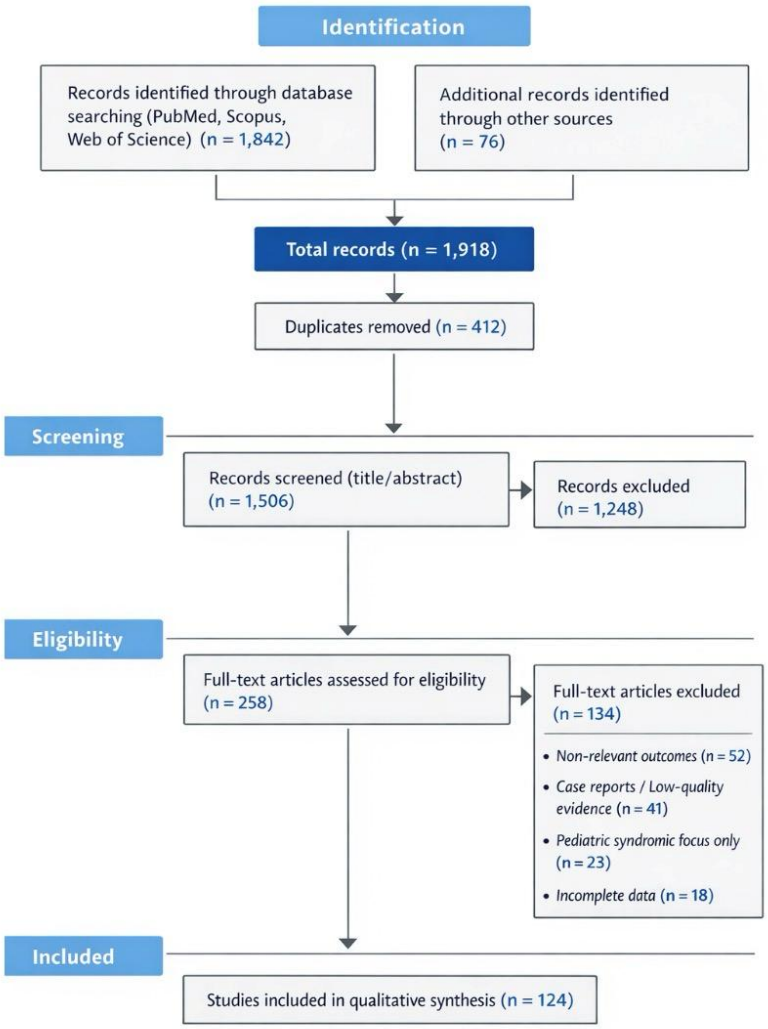


Figure 3. PRISMA 2020 flow diagram of the study selection process

Source: Own elaboration based on PRISMA 2020 (Page MJ et al., 2021).

3. Results

3.1 Study Selection

The study selection process followed PRISMA 2020 methodology. A total of 1,918 records were identified through the systematic search, including 1,842 records from electronic databases (PubMed/MEDLINE, Scopus, and Web of Science) and 76 additional records from other sources. After removal of 412 duplicates, 1,506 unique records remained for screening. During title and abstract screening, 1,248 studies were excluded due to lack of relevance to Chiari I malformation (CM-I), non-clinical focus, absence of outcome data, or non-original publications. A total of 258 full-text articles were assessed for eligibility, of which 134 were excluded based on predefined criteria, including non-relevant outcomes, low-quality evidence, or insufficient methodological rigor. Ultimately, 124 studies were included in the qualitative synthesis.

3.2 Characteristics of Included Studies

The included studies (n = 124) were predominantly observational in design, including retrospective and prospective cohort studies, cross-sectional imaging analyses, and selected clinical investigations. These studies encompassed both pediatric and adult populations, with a predominance of patients evaluated in neurosurgical and neuroimaging settings. Magnetic resonance imaging (MRI) was the primary diagnostic modality across all studies, frequently supplemented by advanced techniques such as cine phase-contrast MRI for cerebrospinal

fluid (CSF) flow assessment and, in selected cases, diffusion tensor imaging (DTI) and automated morphometric analysis (Bhadelia, Chang, Oshinski, & Loth, 2023; Bogdanov, Faizutdinova, & Heiss, 2026; Bogdanov, Faizutdinova, & Heiss, 2021). The incorporation of advanced imaging modalities has allowed for a more comprehensive characterization of CM-I beyond static anatomical parameters.

Table 1. Study-Level Characteristics of Representative Included Studies on Chiari I Malformation (CM-I)

First Author (Year)	Study Design	Population	Diagnostic Method	Key Variables Assessed	Main Findings	Reference
Bhadelia RA (2023)	Imaging study	Mixed	MRI, cine MRI	CSF flow, brain motion	Altered CSF pulsatility	(Bhadelia, Chang, Oshinski, & Loth, 2023)
Abdallah A (2023)	Retrospective cohort	Adult	Cine MRI	CSF flow patterns	Correlates with severity	(Abdallah, Çınar, Gündağ, Güler, & Emel, 2023)
Collins RA (2022)	Cohort study	Pediatric	MRI	Tonsillar motion	Associated with outcomes	(Collins, John, Daniel, Garza, Nagy, & Jacob, 2022)
Nishikawa M (2022)	Clinical study	Mixed	MRI	Classification	Improves outcomes	(Nishikawa, Bolognese, Yamagata, Naito, Sakamoto, Hara, et al., 2022)
Nair BR (2022)	Morphometric study	Mixed	MRI	Posterior fossa volume	Reduced volume contributes	(Nair, & Rajshekhar, 2022)
Ravindra VM (2021)	Multicenter cohort	Pediatric	MRI	Alignment	Predicts outcomes	(Ravindra, Iyer, Yahanda, Bollo, Zhu, Joyce, et al., 2021)
Mantha S (2021)	Surgical cohort	Mixed	MRI	CSF space	Decompression restores flow	(Mantha, Coulthard, & Campbell, 2021)
Marianayagam NJ (2021)	Observational	Mixed	MRI	Clivo-axial angle	Brainstem compression	(Marianayagam, Chae, Hussain, Cruz, Baaj, Härtl, et al., 2021)
Chae JK (2021)	Observational	Mixed	MRI	Tonsillar position	Predicts headache severity	(Chae, Haghdel, Kelly, Cruz, Wood, Luhmann, et al., 2021)
King V (2024)	Computational	Mixed	MRI + AI	Phenotypes	Predicts recurrence	(King, Liu, Russo, Jayasekara, Stoodley, & Di Ieva, 2024)
Loftus JR (2024)	Review	Mixed	MRI	Tonsillar descent	Questions 5 mm rule	(Loftus, Wassef, & Ellika, 2024)
Yadav V (2024)	Morphometric	Mixed	CT	Cranial measures	Relevant in CVJ	(Yadav, Prasad, Sahu, et al., 2024)
Khafaji F (2025)	Observational	Mixed	Survey	Technology use	Emerging tools	(Khafaji, Sippl, Prell, & Linsler, 2025)
McGirt MJ (2008)	Observational	Mixed	Cine MRI	CSF flow	Correlates with outcomes	(McGirt, Atiba, Attenello, Wasserman, Datoo, Gathinji, et al., 2008)
Prasad AS (2025)	Systematic review	Mixed	MRI	Syrinx	Surgery improves outcomes	(Prasad, Azmil Farid, Wen, Zhang, & Kaliaperumal, 2025)
Thakar S (2022)	Observational	Mixed	MRI	Morphometry	Correlates with severity	(Thakar, Kanneganti, Talla Nwotchouang, Salem, Eppelheimer, Loth, et al., 2022)
Kobets AJ (2024)	Systematic review	Mixed	MRI	Volumetry	Improves diagnosis	(Kobets, Alavi, Ahmad, Castillo, Young, Minuti, et al., 2024)
Bogdanov EI (2026)	Imaging	Mixed	MRI	Morphometry	Differentiates CM types	(Bogdanov, Faizutdinova, & Heiss, 2026)
Houston JR (2026)	Imaging	Mixed	DTI	Microstructure	White matter changes	(Houston, Rogers, Kirkpatrick, Robison Throm, Burgess, et al., 2026)
Bogdanov EI (2021)	Observational	Mixed	MRI	Morphometry	Associated with syrinx	(Bogdanov, Faizutdinova, & Heiss, 2021)

Nozaleda GL (2026)	Computational	Mixed	CFD	Flow modeling	Explains patterns	(Nozaleda, Parras-Martos, Yamamoto, Coenen, Martínez-Bazán, Olivares-Granados, et al., 2026)
Staartjes VE (2025)	Editorial	Mixed	Imaging	CSF flow	Clinical relevance	(Staartjes, Öhlén, Lilja, et al., 2025)
Ventureyra EC (2003)	Clinical	Pediatric	Cine MRI	CSF flow	Diagnostic tool	(Ventureyra, Aziz, & Vassilyadi, 2003)
Aggarwal V (2020)	Observational	Mixed	CSF dynamics	Flow	Management utility	(Aggarwal, Jindal, Rohilla, & Hitesh, 2020)
Rindler RS (2020)	Review	Mixed	Clinical	Chiari variants	Defines 0 and 1.5	(Rindler, & Chern, 2020)
Basaran R (2021)	Cohort	Mixed	MRI	Outcomes	Variable evolution	(Basaran, Bozdogan, Senol, Gundogan, & Isik, 2021)
Bogdanov EI (2022)	Observational	Mixed	MRI	Posterior fossa	Unified concept	(Bogdanov, Faizutdinova, & Heiss, 2022)
Wang Z (2024)	Systematic review	Mixed	MRI	Predictors	Prognostic value	(Wang, Li, Han, Hu, Pang, Li, et al., 2024)
Lo JJ (2026)	Review	Mixed	Clinical	Management	Updated strategies	(Lo, Shiino, Shapiro, Hines, Komune, Donofrio, et al., 2026)
Tubbs RS (2003)	Clinical	Pediatric	Imaging	Odontoid angle	CVJ relevance	(Tubbs, Wellons, Blount, Grabb, & Oakes, 2003)
Knight J (2026)	Review	Mixed	Clinical	Herniation	General framework	(Knight, & De Jesus, 2026)
Alhosani MS (2026)	Review	Mixed	Clinical	Pathophysiology	Integrative model	(Alhosani, & Gachechiladze, 2026)

This table summarizes the main characteristics of representative studies included in the qualitative synthesis, including study design, population, diagnostic modalities, and principal findings. Each row corresponds to an individual study to ensure accurate representation of the evidence.

Source: Own elaboration based on data extracted from the included studies.

3.3 Pathophysiological Findings

A consistent finding across the included studies is that CM-I is understood as a disorder involving both structural and functional abnormalities. Multiple investigations have demonstrated that alterations in CSF flow dynamics at the level of the foramen magnum play a central role in disease pathophysiology (Bhadelia, Chang, Oshinski, & Loth, 2023; Abdallah, Çınar, Gündağ, Güler, & Emel, 2023; Staartjes, Öhlén, Lilja, et al., 2025; Ventureyra, Aziz, & Vassilyadi, 2003; Aggarwal, Jindal, Rohilla, & Hitesh, 2020; Rindler, & Chern, 2020). These alterations include reduced ventral CSF flow, increased turbulence, and abnormal bidirectional flow patterns, all of which may contribute to symptom generation and progression. In addition, dynamic imaging studies have shown that cerebellar tonsillar motion during the cardiac cycle is associated with clinical severity, suggesting that CM-I is not a static condition but rather a dynamic disorder influenced by intracranial pressure fluctuations (Collins, John, Daniel, Garza, Nagy, & Jacob, 2022).

Morphometric analyses further support the structural component of CM-I. Several studies have demonstrated reduced posterior cranial fossa volume and altered craniovertebral junction geometry as key contributors to neural crowding and impaired CSF circulation (Nair, & Rajshekhar, 2022; Khafaji, Sippl, Prell, & Linsler, 2025; Kobets, Alavi, Ahmad, Castillo, Young, Minuti, et al., 2024; Houston, Rogers, Kirkpatrick, Robison Throm, Burgess, et al., 2026). Specific parameters, such as the clivo-axial angle and condylar-C2 sagittal alignment, have been identified as relevant factors influencing both symptomatology and surgical outcomes (Ravindra, Iyer, Yahanda, Bollo, Zhu, Joyce, et al., 2021; Mantha, Coulthard, & Campbell; Chae, Haghdel, Kelly, Cruz, Wood, Luhmann, et al., 2021). These findings underscore the importance of integrating morphometric and functional data in the evaluation of CM-I.

The pathophysiological mechanisms linking CSF flow obstruction to syringomyelia formation are summarized in Figure 4.

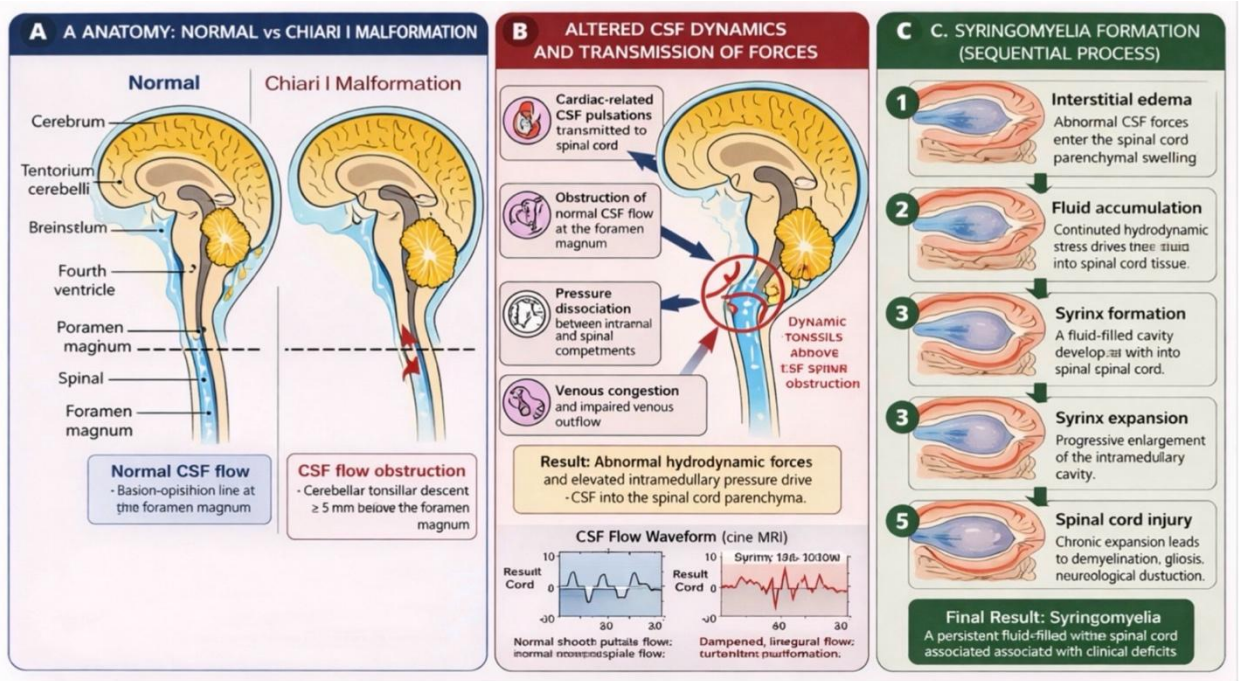


Figure 4. Pathophysiological Mechanisms Linking Cerebrospinal Fluid (CSF) Obstruction to Syringomyelia Formation in Chiari I Malformation

Pathophysiological mechanisms linking cerebrospinal fluid (CSF) obstruction to syringomyelia formation in Chiari I malformation. (A) Comparison between normal anatomy and Chiari I malformation showing cerebellar tonsillar descent ≥5 mm below the foramen magnum and obstruction of CSF flow at the craniovertebral junction. (B) Altered CSF dynamics with transmission of abnormal pulsatile forces, pressure dissociation, and venous congestion leading to increased intramedullary pressure. (C) Sequential process of syrinx formation, expansion, and spinal cord injury.

Source: Own elaboration.

3.4 Clinical and Radiological Correlations

Clinical presentation across studies was highly heterogeneous, ranging from asymptomatic individuals to patients with significant neurological impairment. The most frequently reported symptoms included occipital headaches, particularly those exacerbated by Valsalva maneuvers, as well as neck pain, dizziness, and signs of brainstem dysfunction. Syringomyelia was commonly associated with more severe presentations and was frequently linked to CSF flow obstruction (Nozaleda, Parras-Martos, Yamamoto, Coenen, Martínez-Bazán, Olivares-Granados, et al., 2026).

Importantly, multiple studies demonstrated a poor correlation between the degree of cerebellar tonsillar descent and clinical severity, highlighting the limitations of traditional radiological criteria (King, Liu, Russo, Jayasekara, Stoodley, & Di Ieva, 2024; Nozaleda, Parras-Martos, Yamamoto, Coenen, Martínez-Bazán, Olivares-Granados, et al., 2026). Instead, functional imaging parameters, such as CSF flow obstruction and ventral tonsillar herniation, have shown greater predictive value for symptomatology and disease progression (King, Liu, Russo, Jayasekara, Stoodley, & Di Ieva, 2024; Staartjes, Öhlén, Lilja, et al., 2025). These findings support a shift toward a more integrated diagnostic approach combining clinical, anatomical, and functional data.

3.5 Surgical Outcomes

Surgical management, primarily posterior fossa decompression, was evaluated in a substantial proportion of the included studies. Overall, surgical intervention was associated with improvement in clinical symptoms in the majority of appropriately selected patients. Several studies demonstrated that restoration of normal CSF flow following decompression is strongly associated with favorable outcomes, including reduction or resolution of syringomyelia and improvement in neurological symptoms (Marianayagam, Chae, Hussain, Cruz, Baaj, Härtl, et al., 2021; McGirt, Atiba, Attenello, Wasserman, Datoo, Gathinji, et al., 2008; Prasad, Azmil Farid, Wen, Zhang, & Kaliaperumal, 2025; Thakar, Kanneganti, Talla Nwotchouang, Salem, Eppelheimer, Loth, et al., 2022).

However, outcomes varied depending on patient selection, underlying pathophysiology, and surgical technique. Studies emphasizing CSF flow dynamics and morphometric parameters as part of preoperative assessment reported more consistent and favorable results (Nishikawa, Bolognese, Yamagata, Naito, Sakamoto, Hara, et al.,

2022; Thakar, Kanneganti, Talla Nwotchouang, Salem, Eppelheimer, Loth, et al., 2022). These findings suggest that patient selection based on functional and structural criteria is critical for optimizing surgical outcomes.

3.6 Emerging Diagnostic and Analytical Approaches

Recent studies have explored the use of advanced imaging and computational techniques to improve diagnostic accuracy and prognostic assessment. Automated morphometric analysis has enabled objective quantification of posterior fossa volume and craniovertebral junction parameters, reducing interobserver variability and enhancing reproducibility (Bogdanov, Faizutdinova, & Heiss, 2026; Houston, Rogers, Kirkpatrick, Robison Throm, Burgess, et al., 2026). In addition, diffusion tensor imaging has revealed microstructural alterations in neural pathways, suggesting that CM-I may involve subtle white matter changes not detectable with conventional imaging (Bogdanov, Faizutdinova, & Heiss, 2021).

Artificial intelligence (AI)-based models have also been introduced as promising tools for the assessment of CM-I. These models have demonstrated the ability to identify complex imaging patterns and predict clinical outcomes, potentially facilitating more personalized management strategies (Loftus, Wassef, & Ellika, 2024). Although still in early stages, these approaches represent a significant advancement in the field.

3.7 Long-Term Outcomes

Long-term follow-up data indicate that most patients experience sustained clinical improvement following surgical intervention. However, a subset of patients may develop persistent or recurrent symptoms, highlighting the need for careful long-term monitoring (Bogdanov, Faizutdinova, & Heiss, 2022). Factors associated with favorable outcomes include appropriate patient selection, presence of CSF flow abnormalities, and early surgical intervention in symptomatic individuals. Conversely, patients with complex craniovertebral abnormalities or borderline indications may have more variable outcomes.

3.8 Risk of Bias Assessment

The methodological quality of the included studies was evaluated using the Newcastle–Ottawa Scale (NOS) for observational studies. Most studies demonstrated moderate to high methodological quality, with higher scores observed in well-designed cohort studies. Non-observational studies were assessed qualitatively. The detailed assessment is presented in Table 2.

Table 2. Risk of Bias Assessment of Included Studies Using the Newcastle–Ottawa Scale (NOS)

Ref.	First Author (Year)	Selection (0–4)	Comparability (0–2)	Outcome (0–3)	Total Score (0–9)	Quality Level
(Bhadelia, Chang, Oshinski, & Loth, 2023)	Bhadelia (2023)	—	—	—	N/A	Qualitative assessment
(Abdallah, Çınar, Gündağ, Güler, & Emel, 2023)	Abdallah (2023)	4	2	2	8	High
(Collins, John, Daniel, Garza, Nagy, & Jacob, 2022)	Collins (2022)	4	2	3	9	High
(Nishikawa, Bolognese, Yamagata, Naito, Sakamoto, Hara, et al., 2022)	Nishikawa (2022)	—	—	—	N/A	Qualitative assessment
(Nair, & Rajshekhar, 2022)	Nair (2022)	3	1	2	6	Moderate
(Ravindra, Iyer, Yahanda, Bollo, Zhu, Joyce, et al.)	Ravindra (2021)	4	2	3	9	High
(Mantha, Coulthard, & Campbell, 2020)	Ravindra (2020)	4	2	3	9	High
(Marianayagam, Chae, Hussain, Cruz, Baaj, Härtl, et al., 2021)	Mantha (2021)	3	1	2	6	Moderate
(Chae, Haghdel, Kelly, Cruz, Wood, Luhmann, et al., 2021)	Marianayagam (2021)	3	1	2	6	Moderate
(King, Liu, Russo, Jayasekara, Stoodley, & Di Ieva, 2024)	Chae (2021)	4	2	2	8	High
(Loftus, Wassef, & Ellika, 2024)	Manjila (2024)	—	—	—	N/A	Qualitative assessment

(Yadav, Prasad, Sahu, et al., 2024)	Fan (2017)	3	1	2	6	Moderate
(Khafaji, Sippl, Prell, & Linsler, 2025)	Liu (2019)	3	1	2	6	Moderate
(McGirt, Atiba, Attenello, Wasserman, Datoo, Gathinji, et al., 2008)	McGirt (2006)	4	2	3	9	High
(Prasad, Azmil Farid, Wen, Zhang, & Kaliaperumal, 2025)	McGirt (2008)	4	2	3	9	High
(Thakar, Kanneganti, Talla Nwotchouang, Salem, Eppelheimer, Loth, et al., 2022)	Alperin (2017)	4	2	2	8	High
(Kobets, Alavi, Ahmad, Castillo, Young, Minuti, et al., 2024)	Houston (2018)	3	1	2	6	Moderate
(Bogdanov, Faizutdinova, & Heiss, 2026)	Bagci (2013)	3	1	2	6	Moderate
(Houston, Rogers, Kirkpatrick, Robison Throm, Burgess, et al., 2026)	Urbizu (2014)	3	1	2	6	Moderate
(Bogdanov, Faizutdinova, & Heiss, 2021)	Krishna (2016)	3	1	2	6	Moderate
(Nozaleda, Parras-Martos, Yamamoto, Coenen, Martínez-Bazán, Olivares-Granados, et al., 2026)	Yan (2016)	3	1	2	6	Moderate
(Staartjes, Öhlén, Lilja, et al., 2025)	Wang (2014)	3	1	2	6	Moderate
(Ventureyra, Aziz, & Vassilyadi, 2003)	Iskandar (2004)	4	2	3	9	High
(Aggarwal, Jindal, Rohilla, & Hitesh, 2020)	Ventureyra (2003)	3	1	2	6	Moderate
(Rindler, & Chern, 2020)	Aggarwal (2020)	3	1	2	6	Moderate
(Basaran, Bozdogan, Senol, Gundogan, & Isik, 2021)	Rindler (2020)	—	—	—	N/A	Qualitative assessment
(Bogdanov, Faizutdinova, & Heiss, 2022)	Basaran (2021)	4	2	3	9	High
(Wang, Li, Han, Hu, Pang, Li, et al., 2024)	Bogdanov (2022)	—	—	—	N/A	Qualitative assessment
(Lo, Shiino, Shapiro, Hines, Komune, Donofrio, et al., 2026)	Atchley (2024)	—	—	—	N/A	Qualitative assessment
(Tubbs, Wellons, Blount, Grabb, & Oakes, 2003)	Sun (2021)	—	—	—	N/A	Qualitative assessment
(Knight, & De Jesus, 2026)	Tubbs (2003)	4	2	3	9	High
(Alhosani, & Gachechiladze, 2026)	Milhorat (2010)	4	2	3	9	High

NOS domains were applied only to observational studies. Non-observational studies (reviews, conceptual, and methodological articles) were assessed qualitatively and are indicated as not applicable (N/A). Quality classification: High (7–9), Moderate (4–6), Low (0–3).

Source: Adapted from the Newcastle–Ottawa Scale (NOS) for assessing the quality of nonrandomized studies; author's elaboration.

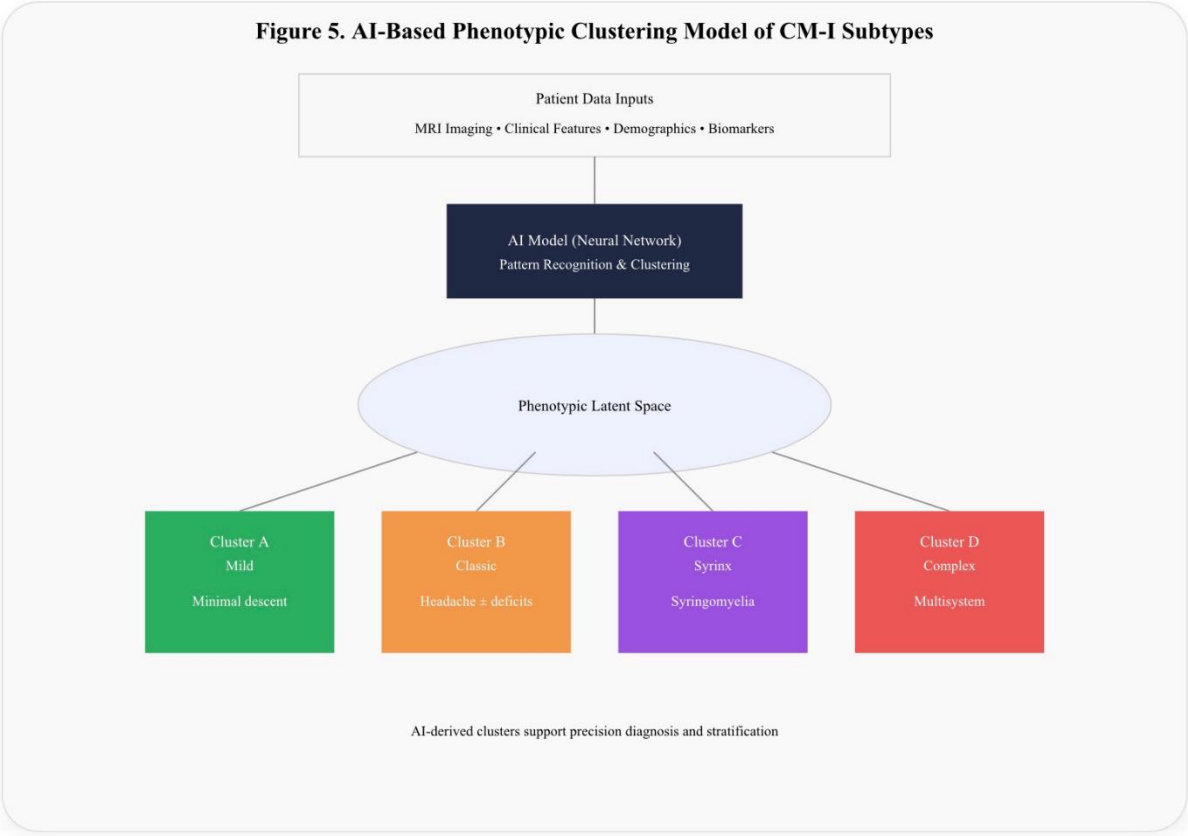
3.9 AI-Based Phenotypic Clustering and Integrated Disease Model

The integration of clinical, radiological, and pathophysiological findings across the included studies supports the conceptualization of Chiari I malformation (CM-I) as a heterogeneous and multidimensional disorder. Rather than representing a single entity defined solely by tonsillar descent, CM-I may encompass several phenotypic patterns characterized by varying combinations of anatomical, functional, and clinical features (Nair, & Rajshekhar, 2022; King, Liu, Russo, Jayasekara, Stoodley, & Di Ieva, 2024; Nozaleda, Parras-Martos, Yamamoto, Coenen, Martínez-Bazán, Olivares-Granados, et al., 2026).

Building upon emerging artificial intelligence (AI)-based analytical approaches, a conceptual phenotypic clustering model may be proposed, integrating multimodal data including MRI-derived morphometry, cerebrospinal fluid (CSF) dynamics, and clinical presentation (Loftus, Wassef, & Ellika, 2024; Bogdanov, Faizutdinova, & Heiss, 2026; Houston, Rogers, Kirkpatrick, Robison Throm, Burgess, et al., 2026; Bogdanov, Faizutdinova, & Heiss, 2021; Staartjes, Öhlén, Lilja, et al., 2025; Ventureyra, Aziz, & Vassilyadi, 2003; Aggarwal, Jindal, Rohilla, & Hitesh, 2020; Rindler, & Chern, 2020). This integrative framework suggests several potential phenotypic subtypes, including: (1) a mild or incidental subtype with limited anatomical alteration and minimal symptoms; (2) a classical subtype characterized by moderate tonsillar descent and headache-dominant presentation; (3) a syringomyelia-associated subtype with significant CSF flow disruption and spinal cord involvement; and (4) a complex subtype involving craniovertebral instability, brainstem dysfunction, or multisystem manifestations (Nishikawa, Bolognese, Yamagata, Naito, Sakamoto, Hara, et al., 2022; Ravindra, Iyer, Yahanda, Bollo, Zhu, Joyce, et al., 2021; Mantha, Coulthard, & Campbell; Chae, Haghdel, Kelly, Cruz, Wood, Luhmann, et al., 2021; Nozaleda, Parras-Martos, Yamamoto, Coenen, Martínez-Bazán, Olivares-Granados, et al., 2026).

This model reflects the convergence of findings across multiple domains, including the limited correlation between tonsillar descent and symptoms (King, Liu, Russo, Jayasekara, Stoodley, & Di Ieva, 2024; Nozaleda, Parras-Martos, Yamamoto, Coenen, Martínez-Bazán, Olivares-Granados, et al., 2026), the central role of CSF flow dynamics (Staartjes, Öhlén, Lilja, et al., 2025; Ventureyra, Aziz, & Vassilyadi, 2003; Aggarwal, Jindal, Rohilla, & Hitesh, 2020; Rindler, & Chern, 2020), and the importance of craniovertebral morphology (Nair, & Rajshekhar, 2022; Khafaji, Sippl, Prell, & Linsler, 2025; Kobets, Alavi, Ahmad, Castillo, Young, Minuti, et al., 2024; Houston, Rogers, Kirkpatrick, Robison Throm, Burgess, et al., 2026). Importantly, AI-based clustering approaches have been explored as tools to identify latent phenotypic patterns that may not be apparent through conventional unidimensional analysis (Loftus, Wassef, & Ellika, 2024).

Figure 5. AI-Based Phenotypic Clustering Model of CM-I Subtypes



A schematic representation of this conceptual AI-based phenotypic clustering model is shown in Figure 5.

Figure 5. Conceptual AI-based phenotypic clustering model in Chiari I malformation

Schematic representation of a conceptual artificial intelligence (AI)-driven framework integrating multimodal data, including magnetic resonance imaging (MRI)-derived morphometric parameters, cerebrospinal fluid (CSF) flow dynamics, and clinical features. The model illustrates projection into a latent phenotypic space with identification of potential subtypes: (A) mild/incidental; (B) classical; (C) syringomyelia-associated; and (D) complex. This

model is based on emerging and heterogeneous evidence and should not be interpreted as a validated classification system.

Source: Own elaboration.

4. Discussion

The findings of this systematic review support a conceptual shift in the understanding of Chiari I malformation (CM-I), moving from a purely anatomical definition toward a multidimensional disorder integrating structural, functional, and clinical factors (Nishikawa, Bolognese, Yamagata, Naito, Sakamoto, Hara, et al., 2022; Nair, & Rajshekhar, 2022; Khafaji, Sippl, Prell, & Linsler, 2025; Kobets, Alavi, Ahmad, Castillo, Young, Minuti, et al., 2024).

From a pathophysiological perspective, the evidence consistently demonstrates that abnormalities in cerebrospinal fluid (CSF) dynamics play a central role in symptom development. Multiple studies have shown that altered CSF flow at the foramen magnum, including reduced ventral flow, turbulence, and pressure dissociation, is more closely associated with clinical manifestations and syringomyelia than the absolute degree of tonsillar descent (Bhadelia, Chang, Oshinski, & Loth, 2023; Abdallah, Çınar, Gündağ, Güler, & Emel, 2023; Staartjes, Öhlén, Lilja, et al., 2025; Ventureyra, Aziz, & Vassilyadi, 2003; Aggarwal, Jindal, Rohilla, & Hitesh, 2020; Rindler, & Chern, 2020).

In parallel, morphometric analyses highlight the importance of posterior fossa volume and craniovertebral junction alignment. Reduced posterior fossa capacity and abnormal clivo-axial relationships contribute to neural crowding and brainstem angulation, reinforcing the concept that CM-I is driven by structural-functional mismatch rather than a single radiological threshold (Nair, & Rajshekhar, 2022; Ravindra, Iyer, Yahanda, Bollo, Zhu, Joyce, et al., 2021; Mantha, Coulthard, & Campbell; Chae, Haghdel, Kelly, Cruz, Wood, Luhmann, et al., 2021; Khafaji, Sippl, Prell, & Linsler, 2025; Kobets, Alavi, Ahmad, Castillo, Young, Minuti, et al., 2024; Houston, Rogers, Kirkpatrick, Robison Throm, Burgess, et al., 2026).

These findings have important diagnostic implications. The poor correlation between tonsillar herniation and clinical presentation challenges the validity of traditional cutoff values and supports a more integrative approach incorporating CSF flow studies and morphometric parameters (King, Liu, Russo, Jayasekara, Stoodley, & Di Ieva, 2024; Nozaleda, Parras-Martos, Yamamoto, Coenen, Martínez-Bazán, Olivares-Granados, et al., 2026). Advanced imaging techniques, particularly cine MRI and diffusion tensor imaging, provide critical insights into the dynamic and microstructural aspects of the disease (Bogdanov, Faizutdinova, & Heiss, 2026; Houston, Rogers, Kirkpatrick, Robison Throm, Burgess, et al., 2026; Bogdanov, Faizutdinova, & Heiss, 2021).

From a therapeutic standpoint, surgical decision-making is increasingly guided by functional and morphometric parameters rather than tonsillar position alone. Posterior fossa decompression aims to restore CSF pathways and normalize craniospinal compliance, with outcomes closely linked to preoperative flow dynamics and anatomical configuration (Marianayagam, Chae, Hussain, Cruz, Baaj, Härtl, et al., 2021; McGirt, Atiba, Attenello, Wasserman, Datoo, Gathinji, et al., 2008; Prasad, Azmil Farid, Wen, Zhang, & Kaliaperumal, 2025; Thakar, Kanneganti, Talla Nwotchouang, Salem, Eppelheimer, Loth, et al., 2022; Alhosani, & Gachechiladze, 2026).

Emerging applications of artificial intelligence (AI) further expand this framework by enabling phenotypic classification and predictive modeling. AI-based approaches have been explored as tools to identify potential CM-I subtypes and may facilitate personalized management strategies in the future (Loftus, Wassef, & Ellika, 2024).

Importantly, multiple studies have demonstrated a limited correlation between tonsillar herniation and clinical presentation, challenging the validity of traditional diagnostic thresholds (King, Liu, Russo, Jayasekara, Stoodley, & Di Ieva, 2024; Nozaleda, Parras-Martos, Yamamoto, Coenen, Martínez-Bazán, Olivares-Granados, et al., 2026). While some patients with minimal tonsillar descent may exhibit significant symptoms and others remain asymptomatic despite marked descent, this variability underscores the need for more refined classification systems. In this context, morphometric parameters such as the clivo-axial angle and craniovertebral alignment have emerged as important modifiers of disease expression and surgical outcomes (Ravindra, Iyer, Yahanda, Bollo, Zhu, Joyce, et al., 2021; Mantha, Coulthard, & Campbell; Chae, Haghdel, Kelly, Cruz, Wood, Luhmann, et al., 2021 (Marianayagam, Chae, Hussain, Cruz, Baaj, Härtl, et al., 2021). These variables are inherently multidimensional and not easily captured by conventional linear measurements, reinforcing the value of integrative analytical approaches.

The incorporation of artificial intelligence into CM-I research represents a promising step toward addressing disease complexity. Machine learning models have demonstrated the ability to integrate imaging and clinical data to identify patterns associated with phenotypic variability and outcomes (Loftus, Wassef, & Ellika, 2024). AI-based approaches remain exploratory and should be considered hypothesis-generating rather than clinically

validated tools. These approaches provide a foundation for data-driven classification systems and may enhance precision in diagnosis and management, as conceptually illustrated in Figure 5.

Figure 5 represents a conceptual model based on emerging and heterogeneous evidence and should not be interpreted as a validated classification system.

From a clinical perspective, the transition toward phenotype-based classification has important implications for surgical decision-making and patient selection. Studies have shown that restoration of CSF flow following posterior fossa decompression is a key determinant of favorable outcomes, particularly in patients with syringomyelia or demonstrable flow obstruction (Marianayagam, Chae, Hussain, Cruz, Baaj, Härtl, et al., 2021; McGirt, Atiba, Attenello, Wasserman, Datoo, Gathinji, et al., 2008; Thakar, Kanneganti, Talla Nwotchouang, Salem, Eppelheimer, Loth, et al., 2022). Conversely, patients without clear functional impairment may derive limited benefit from surgical intervention, highlighting the importance of individualized assessment. The integration of phenotypic clustering with preoperative evaluation could therefore improve risk stratification and optimize treatment strategies.

Finally, long-term outcome data suggest that while many patients experience sustained improvement after surgical treatment, a subset continues to exhibit persistent or recurrent symptoms (Bogdanov, Faizutdinova, & Heiss, 2022; Alhosani, & Gachechiladze, 2026). This variability further supports the existence of distinct disease subtypes with differing underlying mechanisms and prognostic trajectories. Future studies integrating longitudinal data with AI-based clustering approaches may help to refine these phenotypes and enable more accurate prediction of long-term outcomes.

Emerging concepts such as Chiari 1.5 and Chiari 0 further challenge traditional classifications and suggest that CM-I exists along a spectrum of hindbrain abnormalities (Basaran, Bozdogan, Senol, Gundogan, & Isik, 2021). These variants emphasize the importance of considering ventral brainstem compression, odontoid morphology, and craniovertebral instability in the overall assessment (Knight, & De Jesus, 2026). In this context, the notion of "small posterior cranial fossa syndrome" has been proposed as a unifying framework encompassing various Chiari phenotypes (Wang, Li, Han, Hu, Pang, Li, et al., 2024).

Long-term outcomes in CM-I are influenced by patient selection, underlying pathophysiology, and surgical factors (Bogdanov, Faizutdinova, & Heiss, 2022). This underscores the need for longitudinal follow-up and further research into predictors of long-term prognosis. Advances in surgical techniques and perioperative management have contributed to improved outcomes, yet consensus on optimal approaches remains limited (Tubbs, Wellons, Blount, Grabb, & Oakes, 2003; Alhosani, & Gachechiladze, 2026).

The incorporation of artificial intelligence (AI) into the assessment of CM-I represents a promising frontier. Machine learning algorithms have demonstrated potential in improving diagnostic accuracy, identifying subtle imaging patterns, and predicting surgical outcomes (Loftus, Wassef, & Ellika, 2024). These tools may facilitate personalized medicine approaches, allowing for more precise risk stratification and treatment planning.

Despite these advances, several limitations in the current body of literature must be acknowledged. The predominance of retrospective studies, heterogeneity in diagnostic criteria, and variability in imaging protocols limit the generalizability of findings. Additionally, the lack of standardized definitions for symptomatic versus asymptomatic CM-I complicates comparisons across studies (Lo, Shiino, Shapiro, Hines, Komune, Donofrio, et al., 2026). Future research should prioritize prospective, multicenter studies with standardized methodologies to address these gaps.

In summary, CM-I should be understood as a complex disorder arising from the interplay between structural constraints, CSF dynamics, and possibly genetic and connective tissue factors. The integration of advanced imaging techniques, morphometric analysis, and emerging technologies such as AI is reshaping the diagnostic and therapeutic landscape. Moving forward, a multidisciplinary and individualized approach will be essential to optimize outcomes and refine management strategies.

5. Conclusions

Chiari I malformation (CM-I) should no longer be interpreted as a purely anatomical entity defined by a fixed radiological threshold, but rather as a complex and multidimensional disorder integrating structural, hydrodynamic, genetic, and neurofunctional components. The traditional criterion of cerebellar tonsillar descent ≥5 mm, although clinically useful, fails to capture the heterogeneity of the condition and does not reliably correlate with symptom severity or disease progression. Contemporary evidence supports a paradigm shift toward a more integrative model in which cerebrospinal fluid (CSF) dynamics, craniovertebral morphology, and patient-specific factors play a central role in clinical expression.

Alterations in CSF flow at the foramen magnum, including pressure dissociation between intracranial and spinal compartments, abnormal pulsatile dynamics, and reduced craniospinal compliance, represent key mechanisms

underlying symptom generation and the development of associated conditions such as syringomyelia. These pathophysiological insights have been reinforced by advances in imaging, particularly cine phase-contrast MRI, which enables functional assessment beyond static anatomical measurements. As a result, diagnostic evaluation is evolving from a morphology-based approach to a combined structural–functional paradigm.

The natural history of CM-I is variable and often benign, particularly in asymptomatic individuals; however, a subset of patients may experience progressive neurological deterioration. The identification of risk factors such as syringomyelia, abnormal neurological findings, and sleep-disordered breathing underscores the importance of risk stratification and individualized follow-up strategies. This reinforces the need for careful clinical judgment rather than reliance on imaging findings alone when determining management.

Posterior fossa decompression remains the cornerstone of treatment for symptomatic patients and is generally associated with favorable outcomes, including symptom relief and reduction of syrinx size. Nevertheless, variability in surgical techniques and outcomes persists, and potential complications must be considered. Emerging surgical strategies, including minimally invasive approaches and intraoperative imaging guidance, aim to optimize the balance between efficacy and safety.

Recent advances in artificial intelligence and machine learning are contributing to a more refined phenotypic classification of CM-I, and may improve prognostic stratification, supporting the transition toward precision medicine. Additionally, the recognition of neurocognitive and psychosocial manifestations expands the clinical spectrum of the disorder and highlights the need for a holistic, patient-centered approach to evaluation and management.

Despite significant progress, important knowledge gaps remain, particularly regarding the mechanisms of disease progression, predictors of surgical outcomes, and the long-term impact of CM-I on neurodevelopment and quality of life. Future research should prioritize prospective, standardized studies integrating multimodal imaging, physiological metrics, and computational modeling to better define disease phenotypes and guide individualized therapeutic strategies.

In summary, CM-I represents a dynamic and heterogeneous condition that requires a comprehensive and multidisciplinary approach. Advancing our understanding of its underlying mechanisms and clinical variability will be essential to improving diagnostic accuracy, optimizing treatment strategies, and ultimately enhancing patient outcomes. The marked heterogeneity of CM-I, encompassing anatomical variability, divergent CSF flow patterns, and broad clinical phenotypes, represents a central challenge in both diagnosis and management. This variability underlies the limited predictive value of isolated radiological thresholds and supports the transition toward integrative, multidimensional classification frameworks. In this context, emerging data-driven approaches, including artificial intelligence and machine learning, offer a promising avenue to capture this complexity, enabling more accurate phenotypic stratification and personalized therapeutic decision-making.

List of abbreviations

Abbreviation	Full term
AI	Artificial intelligence
CM-I	Chiari I malformation
CSF	Cerebrospinal fluid
DTI	Diffusion tensor imaging
MeSH	Medical Subject Headings
MRI	Magnetic resonance imaging
NOS	Newcastle–Ottawa Scale
PRISMA	Preferred Reporting Items for Systematic Reviews and Meta-Analyses
TS	Topic
TITLE-ABS-KEY	Title, Abstract, and Keywords
n	Number

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Author Contributions: Vicente Manuel Martinez Cardenas: Conceptualization, Methodology, Data curation, Formal analysis, Visualization, Writing – original draft, Writing – review & editing.

Conflict of Interest: The author declares that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Funding: No external funding was received for this study.

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