Original Article

Predictors of reoperation for spinal disorders in Chiari malformation patients with prior surgical decompression

ABSTRACT

Background: Chiari malformation (CM) is a cluster of related developmental anomalies of the posterior fossa ranging from asymptomatic to fatal. Cranial and spinal decompression can help alleviate symptoms of increased cerebrospinal fluid pressure and correct spinal deformity. As surgical intervention for CM increases in frequency, understanding predictors of reoperation may help optimize neurosurgical planning.

Materials and Methods: This was a retrospective analysis of the prospectively collected Healthcare Cost and Utilization Project's California State Inpatient Database years 2004–2011. Chiari malformation Types 1–4 (queried with ICD-9 CM codes) with associated spinal pathologies undergoing stand-alone spinal decompression (queried with ICD-9 CM procedure codes) were included. Cranial decompressions were excluded.

Results: One thousand four hundred and forty-six patients (29.28 years, 55.6% of females) were included. Fifty-eight patients (4.01%) required reoperation (67 reoperations). Patients aged 40–50 years had the most reoperations (11); however, patients aged 15–20 years had a significantly higher reoperation rate than all other groups (15.5% vs. 8.2%, $P = 0.048$). Female gender was significantly associated with reoperation (67.2% vs. 55.6%, $P = 0.006$). Medical comorbidities associated with reoperation included chronic lung disease (19% vs. 6.9%, $P < 0.001$), iron deficiency anemia (10.3% vs. 4.1%, $P = 0.024$), and renal failure (3.4% vs. 0.9%, $P = 0.05$). Associated significant cluster anomalies included spina bifida (48.3% vs. 34.8%, $P = 0.035$), tethered cord syndrome (6.9% vs. 2.1%, $P = 0.015$), syringomyelia (12.1% vs. 5.9%, $P = 0.054$), hydrocephalus (37.9% vs. 17.7%, $P < 0.001$), scoliosis (13.8% vs. 6.4%, $P = 0.028$), and ventricular septal defect (6.9% vs. 2.3%, $P = 0.026$).

Conclusions: Multiple medical and CM-specific comorbidities were associated with reoperation. Addressing them, where possible, may aid in improving CM surgery outcomes.

Keywords: Chiari malformation, decompression, predictors, reoperation

Oluwatobi O. Onafowokan, Ankita Das, Jamshaid M. Mir, Haddy Alas, Tyler K. Williamson, Kimberly Mcfarland, Jeffrey Varghese[1], Sara Naessig, Bailey Imbo, Lara Passfall, Oscar Krol, Peter Tretiakov, Rachel Joujon-Roche, Pooja Dave, Kevin Moattari, Stephane Owusu-Sarpong, Jordan Lebovic, Shaleen Vira[2], Bassel Diebo[3], Virginie Lafage[4], Peter Gust Passias

Department of Orthopedic and Neurological Surgery, NYU Langone Orthopaedic Hospital, [4]Department of Orthopaedics, Lenox Hill Hospital, Northwell Health, New York, USA, [1]Twin Cities Spine Centre, Minneapolis, MN, [2]Department of Orthopedic Surgery, Banner Health, Phoenix, AZ, [3]Department of Orthopedic Surgery, Warren Alpert School of Medicine, Brown University, RI, USA

Address for correspondence: Dr. Peter Gust Passias, 301 East 17th St., New York 10003, NY, USA.
E-mail: Peter.Passias@nyumc.org

Submitted: 18-Oct-23 **Accepted:** 10-Nov-23
Published: 29-Nov-23

INTRODUCTION

Chiari malformations (CM) denote a spectrum of hindbrain abnormalities, characterized by varying degrees of caudal herniation of the hindbrain structures (cerebellum, pons, and medulla oblongata) through the foramen magnum.[1] These malformations are classified into three types based on the anatomical morphology and severity of the defect.[1-3] Treatment of CMs are largely symptom based and include medical and surgical management.

Operative interventions for CM are considered when affected individuals experience persistent and debilitating

Access this article online	
Website: www.jcvjs.com	Quick Response Code
DOI: 10.4103/jcvjs.jcvjs_140_23	

How to cite this article: Onafowokan OO, Das A, Mir JM, Alas H, Williamson T, Mcfarland K, *et al.* Predictors of reoperation for spinal disorders in Chiari malformation patients with prior surgical decompression. J Craniovert Jun Spine 2023;14:336-40.

symptoms.[4,5] The goals of surgery are to relieve pressure on the relevant hindbrain structures and to restore the normal cerebrospinal fluid (CSF) flow across the craniovertebral junction, which is most commonly done by decompression of the posterior fossa and foramen magnum.[5-7] Reasons for CM reoperation include persisting neurological symptoms, inadequate initial decompression, faulty stent placements, and development of postoperative complications.[8,9]

With the number of CM patients being treated with spinal decompression increasing,, it becomes even more important to investigate which patients may be at risk for poor outcomes.[10] Therefore, it becomes increasingly imperative to understand the risk factors that may lead to revision surgery. To our knowledge, there is very little in the literature on possible predictors for reoperation. Thus, this study aims to investigate the predictors of reoperation in CM patients with previous surgical history of stand-alone spinal decompression.

MATERIALS AND METHODS

Study design and data source

This was a retrospective cohort analysis of prospectively collected data from the Healthcare Cost and Utilization Project's California State Inpatient Database (HCUP-CSID) during the years 2004–2011. The HCUP-CSID was constructed using data files from the California State Department of Health's Statewide Planning and Research Cooperative System. Data elements in the HCUP-CSID include patient demographics, admission and discharge status, total hospital charges, and length of stay, as well as procedural information and diagnoses as per the International Classification of Diseases Ninth Revision-Clinical Modification (ICD-9-CM) codes. Unique patient-specific linkage codes allowed for longitudinal tracking of patients within the study period. The local Institutional Review Board waived the need for informed consent, as the HCUP-CSID is publicly available and de-identified. Discharges and procedures were coded in the ICD-9-CM format.[11]

Inclusion and exclusion criteria

Patients with a diagnosis of CMs of all types were identified. CMs were assessed by Chiari type using ICD-9-CM codes: Type 1 (ICD-9-CM 348.4), Type 2 (741.00, 741.01, 741.02, or 741.03 without another Chiari code), Type 3 (742.0), and Type 4 (742.2). Reoperations, surgical complications, and comorbidities were identified. We included all patients with a Chiari diagnosis from 2004 to 2011 in the database for a total of 1446 patients.

Statistical analysis

All statistical analyses were performed using SPSS version 23.0 (Armonk, NY, USA: IBM), alongside data preparation using Microsoft Excel 2016 MSO (Redmond, WA, USA: Microsoft). Rates of reoperation and associated characteristics were investigated using contingency tables in each CM cohort. The aforementioned were generated by an algorithm, which selected all those associations co-occurring in at least 0.5% of the unweighted population. This cutoff allowed us to avoid over-representation due to heavily weighted data points in our final assessment. Rates of reoperation and co-occurring demographics on the weighted population of discharged CM patients were then determined by re-generating cross-tabulations for the top-paired associations that met this threshold.

Pearson's Chi-squared (χ^2) tests of independence were utilized to explore significant differences in proportions of reoperations between CM types and other demographic factors. $P < 0.05$ was considered statistically significant.

RESULTS

Cohort overview

Overall, 1446 CM patients were isolated in the HCUP-CSID and included in this analysis. The cohort was comprised of 55.6% of female patients, with a mean patient age of 29.9 years. There were 58 patients (4.01%) who underwent reoperation following their initial spinal decompression surgery.

Surgical overview

Of the 1446 included CM patients who were initially treated with stand-alone spinal decompression, 58 (4.01%) patients underwent a total of 67 reoperations. Of these reoperations, 57 (85.1%) included re-decompression and 10 (14.9%) included fusion only.

Comparison of initial and reoperation and nonreoperation patients

While patients between the ages of 40–50 years had the most reoperations of any age group (11 reoperations), patients between the ages of 15–20 years had the highest reoperation rate compared to other age groups (15.5% vs. 8.2%, $P = 0.048$). Furthermore, gender was significantly associated with reoperation, with 67.2% of reoperations being female ($P = 0.006$).

Medical comorbidities associated with reoperation

Medical comorbidities associated with reoperation included chronic lung disease (19% vs. 6.9%, $P < 0.001$), iron deficiency anemia (10.3% vs. 4.1%, $P = 0.024$), and renal failure (3.4% vs. 0.9%, $P = 0.05$) [Table 1]. Other common medical conditions including chronic heart failure, hypertension, obesity, and depression were not significantly related to rates of reoperation (all $P > 0.05$).

Clusters of anomalies associated with reoperation

Congenital clusters of anomalies related to CM were also associated with increased rates of reoperation. This included those primarily associated with the spine, such as spina bifida (48.3% vs. 34.8%, $P = 0.035$), tethered cord syndrome (6.9% vs. 2.1%, $P = 0.015$), syringomyelia (12.1% vs. 5.9%, $P = 0.054$), and scoliosis (13.8% vs. 6.4%, $P = 0.028$) [Table 2]. Other notable congenital anomalies involved pathological conditions of the brain and heart, including hydrocephalus (37.9% vs. 17.7%, $P < 0.001$) and ventricular septal defect (VSD) (6.9% vs. 2.3%, $P = 0.026$). However, other cardiac-related anomalies, such as atrial septal defects, patent ductus arteriosus, and coarctation of the aorta were not significantly associated with reoperation (all $P > 0.05$).

DISCUSSION

CM diagnoses are commonly treated through spinal decompression surgery, and when successful, these interventions provide a marked improvement in clinical outcomes.[12] As the frequency of surgical intervention for CM has increased, so have the rates of perioperative complications and reoperations. The correlation between patient factors and the need for revision surgery following initial spinal decompression surgery is not well understood, though previous studies have suggested that some patient factors may lead to an increased probability that revision surgery will be needed.[9,13,14]

Table 1: Medical comorbidities associated with reoperation

	Reoperation (%)	Nonreoperation (%)	*P*
Chronic lung disease	19	6.9	<0.001
Iron deficiency anemia	10.3	4.1	0.024
Renal failure	3.4	0.9	0.05
Depression	1.7	1.4	0.133
Hypertension	11.9	9.8	0.166
Diabetes	19.4	19.0	0.221
Chronic heart failure	17.9	19.2	0.33

Table 2: Congenital anomalies associated with reoperation

	Reoperation (%)	Nonreoperation (%)	*P*
Spina bifida	48.3	34.8	0.035
Tethered cord syndrome	6.9	2.1	0.015
Syringomyelia	12.1	5.9	0.054
Scoliosis	13.8	6.4	0.028
Hydrocephalus	37.9	17.7	<0.001
Ventricular septal defect	6.9	2.3	0.026
Atrial septal defect	5.5	7.1	0.421
Patent ductus arteriosus	1.4	1.0	0.185

In this study, we examined the demographic and comorbidity parameters as well as rates of reoperation for 1446 CM patients who underwent initial spinal decompression surgery, to study the relationship between patient-specific parameters and rates of reoperation. Overall, our study found that 58 (4.01%) patients underwent a total of 67 reoperations, and that of these reoperations 57 included re-decompression while 10 included fusions only.

Our study found that certain demographic features were associated with increased rates of reoperation for patients with CM. Of interest was the finding that CM patients aged 15–20 years had the highest rate of reoperation compared to other age groups (15.5% vs. 8.2%, $P = 0.048$). As many patients with CM have near normal life expectancy (particularly for the Type I subtype),[15-17] this may indicate that the potential benefits of reoperation in a younger patient may outweigh the risks in providing many years of improved health. The lower rates of reoperation in older patients may indicate that surgeons are less tolerant of the risks associated with reoperation in older patients who are generally less healthy and for whom the revision surgery may provide fewer years of benefit for. Overall, these demographic trends in reoperation rates suggest that it is worth considering how much benefit a patient may receive from reoperation when planning their treatment.

In addition, it was found that various medical comorbidities were associated with increased rates of reoperation in patients with CM. Among these factors include chronic lung disease (19% vs. 6.9%, $P < 0.001$), iron deficiency anemia (10.3% vs. 4.1%, $P = 0.024$), and renal failure (3.4% vs. 0.9%, $P = 0.05$). This suggests that it is necessary to consider what medical comorbidities a patient has in determining whether they should be candidates for surgery.

Perhaps most interesting are the clusters of anomalies associated with increased rates of reoperation as these appear to be associated with syrinx formation and the embryological basis of CM. Our study found that abnormalities associated with syrinx formation including spina bifida (48.3% vs. 34.8%, $P = 0.035$), tethered cord syndrome (6.9% vs. 2.1%, $P = 0.015$), syringomyelia (12.1% vs. 5.9%, $P = 0.054$), scoliosis (13.8% vs. 6.4%, $P = 0.028$), and hydrocephalus (37.9% vs. 17.7%, $P < 0.001$) were associated with higher rates of reoperation. Many theories have been proposed regarding the pathophysiology of syrinx formation in the setting of CM, including intramedullary and extramedullary obstruction to CSF flow at the level of the foramen magnum.[18-20] Worsening obstruction and deformity likely contribute to the spectrum of disease seen

in CM, with higher rates of hydrocephaly and encephalocele with worsening disease. Depending on the degree of underlying posterior fossa hypoplasia, various neurologic associations including spina bifida, tethered cord syndrome, syringomyelia, and hydrocephalus can manifest. Our study also found that scoliosis is associated with increased rates of reoperation. Previous research reported that the increased rates of scoliosis in patients with CM may be caused by increased pressure from the expansive syrinx interfering with postural reflexes responsible for trunk musculature causing postural imbalance and musculoskeletal deformity.[21,22] This is supported by studies that have demonstrated improvement in scoliosis following CM decompression surgery.[23,24] Finally, we found increased rates of reoperation among CM patients with VSD (6.9% vs. 2.3%, $P = 0.026$). CM has been described as malformations due to inadequacy of the para-axial mesoderm after neural tube closure, which leads to posterior fossa hypoplasia and the associated symptoms and neurologic dysfunction.[25] It may be hypothesized that with more severe derangement in mesodermal growth and development, other mesodermal derivatives including cardiac tissue may also be affected, resulting in congenital heart defects. These disturbances in mesodermal tissue may indicate that CM patients with ventricular septal involvement have worse disease, helping to explain the increase in rates of reoperation. As CM varies in presentation between patients and often affects multiple organ systems, understanding the clusters of abnormalities associated with increased rates of reoperation can be beneficial in determining which patients are the best candidates for surgery and which may need additional monitoring after initial spinal decompression surgery.

More recently, there have been controversial discussions around managing symptomatic CM with solely atlantoaxial stabilization (with plate and screw fixation). This has stemmed from two proposed theories: that basilar invagination and CM have similar etiologies and the associated syringomyelia is due to C1-2 instability (subtle or apparent), and that cerebellar tonsillar herniation may be a compensatory protective mechanism to buffer the spinal cord from the detrimental effects of atlantoaxial instability.[26,27] Definitions of atlantoaxial instability have varied in the literature but have been accepted as atlantodental intervals on radiographs of at least 3 mm in adults and 5 mm in children, or atlantodental intervals on computed tomography of at least 2 mm in adults.[28-31] Numerous studies have investigated atlantoaxial instability as the pathophysiological basis for CM, but none have been able to definitively support or refute it.[32] The clinical benefit of decompression alone for CM has been proven.[33,34] Therefore, it is still not currently possible to recommend sole atlantoaxial stabilization for the management of symptomatic CM. However, it would be prudent to assess any symptomatic CM patient for atlantoaxial instability coexistence, and subsequently determine if C1-2 stabilization procedures are also warranted.[32,35] Although data on atlantoaxial instability were unavailable to us in this study, further prospective studies examining a potential relationship with reoperation after decompression surgery will be beneficial to improving knowledge in this field.

We acknowledge limitations to this study inherent to the use of a retrospective review of large administrative patient databases. Patients were identified using ICD-9-CM coding definitions, which is a potential source of error if coding inaccuracies or misclassifications occur. These codes are often inputted by medical staff with varying levels of clinical training and oversight, which may lead to underreporting or omission of both diagnoses and their various associations. As such, estimates of coding accuracy have been estimated at approximately 80%.[36,37] Furthermore, the HCUP-CSID is reliant on inpatient data from patients in California which may not be completely representative of the general demographics of CM patients in the United States and does not include data for same-day operations. Nonetheless, to our knowledge, this is the largest retrospective cohort study examining the potential predictive characteristics of reoperation in CM patients following initial spinal decompression surgery.

CONCLUSIONS

There are numerous patient factors associated with increased rates of reoperation in CM patients who undergo initial spinal decompression surgery. We found that female patients aged 15–20 years undergo significantly more reoperations after index decompression compared to other CM patients. We also found that patients with non-CM-related comorbidities including chronic lung disease and anemia have higher rates of reoperation. Finally, patients with CM-specific comorbidities including spina bifida, tethered cord, syringomyelia, hydrocephalus, scoliosis, and VSD also had higher rates of reoperation. In conclusion, understanding the patient factors associated with increased rates of reoperation can be useful in surgical planning as it can help determine which patients are the best surgical candidates and lead to better postoperative care through closer monitoring of patients with CM who are more likely to need reoperation.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Kular S, Cascella M. Chiari I malformation. In: StatPearls. Treasure Island (FL): StatPearls Publishing Copyright © 2022, StatPearls Publishing LLC.; 2022.
2. Hidalgo JA, Tork CA, Varacallo M. Arnold chiari malformation. In: StatPearls. Treasure Island (FL): StatPearls Publishing Copyright © 2022, StatPearls Publishing LLC.; 2022.
3. Hadley DM. The chiari malformations. J Neurol Neurosurg Psychiatry 2002;72 Suppl 2:i38-40.
4. Lin W, Duan G, Xie J, Shao J, Wang Z, Jiao B. Comparison of results between posterior fossa decompression with and without duraplasty for the surgical treatment of chiari malformation type I: A systematic review and meta-analysis. World Neurosurg 2018;110:460-74.e5.
5. Giammattei L, Borsotti F, Parker F, Messerer M. Chiari I malformation: Surgical technique, indications and limits. Acta Neurochir (Wien) 2018;160:213-7.
6. Kristiansson H, Fletcher-Sandersjöö A, Cesarini K, Fransson M, Vlachogiannis P, Burström G, *et al.* Dura management strategies in the surgical treatment of adult chiari type I malformation: A retrospective, multicenter, population-based parallel cohort case series. Oper Neurosurg (Hagerstown) 2022;23:304-11.
7. Pritz MB. Surgical treatment of chiari I malformation: Simplified technique and clinical results. Skull Base 2003;13:173-7.
8. Sacco D, Scott RM. Reoperation for chiari malformations. Pediatr Neurosurg 2003;39:171-8.
9. Mazzola CA, Fried AH. Revision surgery for chiari malformation decompression. Neurosurg Focus 2003;15:E3.
10. Passias PG, Pyne A, Horn SR, Poorman GW, Janjua MB, Vasquez-Montes D, *et al.* Developments in the treatment of chiari type 1 malformations over the past decade. J Spine Surg 2018;4:45-54.
11. Agency for Healthcare Research and Quality Healthcare Cost and Utilization Project (HCUP). Introduction To The Hcup Nationwide Inpatient Sample (NIS). 2011. Available from: https://www.hcup-us.ahrq.gov/db/nation/nis/NIS_Introduction_2011.pdf [Last accessed on 2023 Sep 10].
12. Pattisapu JV, Ackerman LL, Infinger LK, Maher CO, Quinsey C, Rocque BG, *et al.* Congress of neurological surgeons systematic review and evidence-based guidelines for patients with chiari malformation: Surgical interventions. Neurosurgery 2023;93:731-5.
13. Chae JK, Greenfield JP. Revision chiari surgery in young children: Predictors and outcomes. Pediatr Neurosurg 2021;56:529-37.
14. Knafo S, Malcoci M, Morar S, Parker F, Aghakhani N. Surgical management after chiari decompression failure: Craniovertebral junction revision versus shunting strategies. J Clin Med 2022;11:3334.
15. Langridge B, Phillips E, Choi D. Chiari malformation type 1: A systematic review of natural history and conservative management. World Neurosurg 2017;104:213-9.
16. Carey M, Fuell W, Harkey T, Albert GW. Natural history of chiari I malformation in children: A retrospective analysis. Childs Nerv Syst 2021;37:1185-90.
17. Maher CO. Natural history of chiari malformations. In: Tubbs RS, Turgut M, Oakes WJ, editors. The Chiari Malformations. Cham: Springer International Publishing; 2020. p. 275-87.
18. Koyanagi I, Houkin K. Pathogenesis of syringomyelia associated with chiari type 1 malformation: Review of evidences and proposal of a new hypothesis. Neurosurg Rev 2010;33:271-84.
19. Meadows J, Kraut M, Guarnieri M, Haroun RI, Carson BS. Asymptomatic chiari type I malformations identified on magnetic resonance imaging. J Neurosurg 2000;92:920-6.
20. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ. Pathophysiology of syringomyelia associated with chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. J Neurosurg 1994;80:3-15.
21. Huebert HT, MacKinnon WB. Syringomyelia and scoliosis. J Bone Joint Surg Br 1969;51:338-43.
22. Eule JM, Erickson MA, O'Brien MF, Handler M. Chiari I malformation associated with syringomyelia and scoliosis: A twenty-year review of surgical and nonsurgical treatment in a pediatric population. Spine (Phila Pa 1976) 2002;27:1451-5.
23. Strahle JM, Taiwo R, Averill C, Torner J, Gewirtz JI, Shannon CN, *et al.* Radiological and clinical associations with scoliosis outcomes after posterior fossa decompression in patients with chiari malformation and syrinx from the Park-Reeves syringomyelia research consortium. J Neurosurg Pediatr 2020;26:53-9.
24. Brockmeyer D, Gollogly S, Smith JT. Scoliosis associated with chiari 1 malformations: The effect of suboccipital decompression on scoliosis curve progression: A preliminary study. Spine (Phila Pa 1976) 2003;28:2505-9.
25. Hiremath SB, Fitsiori A, Boto J, Torres C, Zakhari N, Dietemann JL, *et al*. The perplexity surrounding chiari malformations – Are we any wiser now? AJNR Am J Neuroradiol 2020;41:1975-81.
26. Goel A. Is atlantoaxial instability the cause of chiari malformation? Outcome analysis of 65 patients treated by atlantoaxial fixation. J Neurosurg Spine 2015;22:116-27.
27. Goel A. Is chiari malformation nature's protective "air-bag"? Is its presence diagnostic of atlantoaxial instability? J Craniovertebr Junction Spine 2014;5:107-9.
28. Rojas CA, Bertozzi JC, Martinez CR, Whitlow J. Reassessment of the craniocervical junction: Normal values on CT. AJNR Am J Neuroradiol 2007;28:1819-23.
29. Passias PG, Wang S, Kozanek M, Wang S, Wang C. Relationship between the alignment of the occipitoaxial and subaxial cervical spine in patients with congenital atlantoxial dislocations. J Spinal Disord Tech 2013;26:15-21.
30. King JE, Brumley MA. What is atlantoaxial instability? Nursing 2005;35:71.
31. White AA 3rd, Panjabi MM. The clinical biomechanics of the occipitoatlantoaxial complex. Orthop Clin North Am 1978;9:867-78.
32. Wagner A, Grassner L, Kögl N, Hartmann S, Thomé C, Wostrack M, *et al.* Chiari malformation type I and basilar invagination originating from atlantoaxial instability: A literature review and critical analysis. Acta Neurochir (Wien) 2020;162:1553-63.
33. Chai Z, Xue X, Fan H, Sun L, Cai H, Ma Y, *et al.* Efficacy of posterior fossa decompression with duraplasty for patients with chiari malformation type I: A systematic review and meta-analysis. World Neurosurg 2018;113:357-65.e1.
34. Joaquim AF, Tedeschi H, Chandra PS. Controversies in the surgical management of congenital craniocervical junction disorders – A critical review. Neurol India 2018;66:1003-15.
35. Chatterjee S, Shivhare P, Verma SG. Chiari malformation and atlantoaxial instability: Problems of co-existence. Childs Nerv Syst 2019;35:1755-61.
36. Burns EM, Rigby E, Mamidanna R, Bottle A, Aylin P, Ziprin P, *et al.* Systematic review of discharge coding accuracy. J Public Health (Oxf) 2012;34:138-48.
37. Gologorsky Y, Knightly JJ, Lu Y, Chi JH, Groff MW. Improving discharge data fidelity for use in large administrative databases. Neurosurg Focus 2014;36:E2.