

# Timing to surgery of Chiari malformation type 1 affects complication types: An analysis of 13,812 patients

## ABSTRACT

**Background:** Chiari malformations (CM) are congenital defects due to hypoplasia of the posterior fossa with cerebellar herniation into the foramen magnum and upper spinal canal. Despite the vast research done on this neurological and structural syndrome, clinical features and management options have not yet conclusively evolved. Quantification of proper treatment planning, can lead to potential perioperative benefits based on diagnoses and days to procedure. This study aims to identify if early operation produces better perioperative outcomes or if there are benefits to delaying CM surgery.

**Aims and Objective:** Assess outcomes for Chiari type I.

**Methods:** The KID database was queried for diagnoses of Chiari Malformation from 2003-2012 by icd9 codes (348.4, 741.0, 742.0, 742.2). Included patients: had complete time to procedure (TTP) data. Patients were stratified into 7 groups by TTP: Same-day as admission (SD), 1-day delay (1D), 2-day delay (2D), 3-day delay (3D), 4-7 days delay (4-7D), 8-14 days delay (8-14D), >14 days delay (>14D). Differences in pre-operative demographics (age/BMI) and perioperative complication rates between patient cohorts were assessed using Pearson's chi-squared tests and T-tests. Surgical details, perioperative complications, length of stay (LOS), total charges, and discharge disposition was compared. Binary logistic regressions determined independent predictors of varying complications (reference: same-day).

**Results:** 13,812 Chiari type I patients were isolated from KID (10.12 ± 6.3, 49.2F%, .063 ± 1.3CCI). CM-1 pts were older (10.12 yrs vs 3.62 yrs) and had a higher Charlson Comorbidity Score (0.62 vs 0.53; all  $P < 0.05$ ). Procedure rates: 27.8% laminectomy, 28.3% decompression, and 2.2% spinal fusion. CM-1 experienced more complications (61.2% vs 37.9%) with the most common being related to the nervous system (2.8%), anemia (2.4%), acute respiratory distress disorder (2.1%), and dysphagia (1.2%). SD was associated with the low length of stay (5.3 days vs 9.5-25.2 days,  $P < 0.001$ ), total hospital charges (\$70,265.44 vs \$90, 945.33-\$269, 193.26,  $P < 0.001$ ) when compared to other TTP groups. Relative to SD, all delay groups had significantly increased odds of developing postoperative complications (1D-OR: 1.29 [1.1-1.6] → 8-14D-OR: 4.77[3.4-6.6]; all  $P < 0.05$ ), more specifically, nervous system (1D-OR: 1.8 [1.2-2.5] → 8-14D-OR: 3.3 [1.8-6.2]; all  $P < 0.05$ ). Sepsis complications were associated with a delay of at least 3D(2.5[1.4-4.6]) while respiratory complications (6.2 [3.1-12.3]) and anemia (2 [1.1-3.5]) were associated with a delay of at least 8-14D (all  $P < 0.05$ ).

**Keywords:** Chiari, complications, timing

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## INTRODUCTION

Chiari malformation type 1 (CM1) is the most common subtype of congenital hindbrain anomalies, which is usually asymptomatic in childhood.<sup>[1,2]</sup> It is estimated to occur in up to 3.5% of the general patient population, with a higher

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proportion of females being affected.<sup>[3,4]</sup> Common presenting symptoms in patients who have CM1 often result from direct compression of the neurological structures, such as occipital headaches, cerebellar signs, myelopathy, and respiratory compromise.<sup>[1,5-7]</sup> The onset of these symptoms, which occur in late childhood to adolescence, results in their incidental discovery upon diagnostic neuroimaging.<sup>[8-11]</sup>

Despite decades of experience and research, the management of CM1 continues to be controversial, with much ongoing debate regarding management, including indications, timing, and procedures.<sup>[12-16]</sup> Although medical management is considered for some, a majority of patients require surgery to decompress the posterior fossa, thereby relieving cerebellar pressure and returning cerebrospinal fluid (CSF) flow.<sup>[2,7,13,14,17-19]</sup> The time to diagnosis from the onset of symptomatology has been reported to take over 40 months, with resultant intervention generally occurring between 7 and 24 months, after diagnosis has been established.<sup>[2,20]</sup> Although there are currently various studies evaluating positive and negative predictors for postoperative outcomes, there is currently a paucity of information regarding the effects of point of intervention from diagnosis to index procedure.<sup>[21,22]</sup>

Due to the length of time usually taken to diagnose CM1, there is already an inevitable delay inherent in the management of this condition. Therefore, the timing of surgical management, specifically with early intervention, may indeed impact peri- and postoperative complications. The aim of our study, therefore, was to utilize a large database to identify the effects of timing of the surgical management of CM1 patients to determine if earlier surgical management would result in more favorable postoperative outcomes or if there may be potential benefits to delayed surgery. To the authors' knowledge, this is the first study investigating complication rates related to the timing of CM1 surgical management.

## MATERIALS AND METHODS

### Data source

The kid inpatient database (KID) is the largest publicly-available all-payer pediatric (age <21 years at admission) inpatient healthcare database in the United States. The Agency for Healthcare Research and Quality's Healthcare Cost and Utilization Project (HCUP) created this KID. KID sampling includes complicated and uncomplicated births, as well as other pediatric inpatient procedures from community, nonrehabilitation hospitals. The KID contains 107 data elements, using the International Classification of Disease,

Ninth Revision, Clinical Modification (ICD-9-CM) format to code all of the diagnoses and procedures. With over 3 million hospital stays per 3-year database, it is designed to allow accurate calculation of medical condition incidences using HCUP-provided trend weights.<sup>[1]</sup> A detailed overview of the KID design is available at <https://www.hcup-us.ahrq.gov/kidoverview.jsp>.

### Patient sample

The KID was queried for patients with E-Codes (ICD-9-CM codes) pertaining to CM1 from 2003 to 2012 (348.4, 741.0, 742.0, and 742.2). All these cases were emergent admissions for neurological deterioration. Patients were then further stratified into seven groups by timing to operation: same day as admission (SD), 1-day delay (1D), 2-day delay (2D), 3-day delay (3D), 4–7-day delay (4–7D), 8–14-day delay (8–14D), and >14-day delay (>14D). Differences in preoperative demographics (age/body mass index) and perioperative complication rates between patient cohorts were assessed using Pearson's Chi-square tests and *t*-tests. Surgical details, perioperative complications, length of stay (LOS), total charges, and discharge disposition were compared. Binary logistic regressions determined independent predictors of varying complications (reference: same day).

### Statistical analysis

Descriptive and univariate analyses identified basic demographics and surgical characteristics for CM1 patients. Once patients were stratified according to their timing to operation, differences in outcomes among the groups were assessed through Pearson's Chi-square tests and *t*-tests when appropriate. Outcomes that were identified were LOS, total hospital charges, and postoperative complications such as sepsis, respiratory complications, and anemia. Binary logistic regressions determined how the delay of surgery independently predicted various complications and outcomes (reference: same day). All statistics were done using SPSS Statistics version 23.0 (IBM Corp., Armonk, NY, USA). A statistical cutoff value of  $P < 0.05$  was considered statistically significant.

## RESULTS

### Demographic overview

A total of 13,812 Chiari type I patients were isolated. By basic demographics, the average age was  $10.12 \pm 6.3$  years, 49.2% were female, and the average Charlson comorbidity index (CCI) was  $0.063 \pm 1.3$ . Patients diagnosed with CM1 were older (10.12 vs. 3.62 years) and had a higher Charlson comorbidity score (0.62 vs. 0.53; all  $P < 0.05$ ) than those who were not diagnosed with CM1. More specifically at presentation, 7.3% of the CM1 patients had cerebrovascular

**Table 1: Length of stay and total hospital charges by timing to operation**

| Outcomes      | Same day    | 1-day delay | 2-day delay  | 3-day delay  | 4-7-day delay | 8-14-day delay | 15+ day delay | P      |
|---------------|-------------|-------------|--------------|--------------|---------------|----------------|---------------|--------|
| LOS           | 5.3         | 9.6         | 9.9          | 12.4         | 17.4          | 25.1           | 50.85         | <0.001 |
| Total charges | \$70,265.44 | \$113,414.7 | \$101,783.78 | \$141,586.74 | \$178,619.01  | \$253,013.94   | \$480,555.10  | <0.001 |

LOS - Length of stay

comorbidities, 11.7% had pulmonary issues, 7.9% had a malignancy, and 6% were plegic.

### Surgical overview

By surgical approach, 1.3% of the CM1 patients received a posterior approach, 0.1% received an anterior approach, and 0.1% received a combined approach. 27.8% of the CM1 patients underwent a laminectomy, 28.3% had a decompression, and 2.2% had a spinal fusion. 50.5% of these cases were elective, 30.2% were an emergency, 16.7% were urgent, and 2% were due to trauma.

### Timing to operation

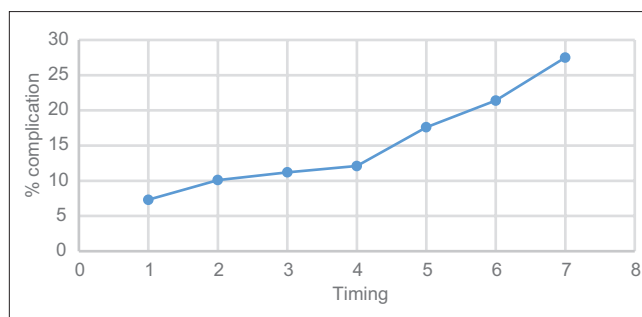
Stratifying by timing to operation, 73.8% received an operation the SD, 10.4% 1D, 4.4% 2D, 3.0% 3D, 4.6% >3D and <8D, 2.2% >8D and <15D, and 1.6% >15D. Patients who received SD surgery were older than the other delayed groups (10.4 vs. 1D: 9.19, 2D: 9.15, 3D: 9.5, 4-7D: 10.0, 8-14D: 8.6, 15D: 9.1;  $P < 0.001$ ). However, those in the 15+ delay group had the greatest CCI (1.6 vs. SD: 0.47, 1D: 1.0, 2D: 1.1, 3D: 1.1, 4-7D: 1.3, 8-14D: 1.4;  $P < 0.001$ ).

### Outcomes by timing to operation

SD was associated with lower LOS [5.3 days vs. 9.5–25.2 days,  $P < 0.001$ ; Table 1]. This group also had lower total hospital charges (\$70,265.44 vs. \$90,945.33–\$269,193.26,  $P < 0.001$ ) when compared to other time to operation groups [Table 1]. Relative to SD, all delay groups had significantly increased odds of developing postoperative complications (1D – odds ratio [OR]: 1.29 [1.1–1.6] → 8-14D – OR: 4.77 [3.4–6.6]; all  $P < 0.05$ ; [Table 2]), more specifically, the nervous system (1D – OR: 1.8 [1.2–2.5] → 8-14D – OR: 3.3 [1.8–6.2]; all  $P < 0.05$ ). Sepsis complications were associated with a delay of at least 3D (2.5 [1.4–4.6]), whereas respiratory complications (6.2 [3.1–12.3]) and anemia (2 [1.1–3.5]) were associated with a delay of at least 8-14D (all  $P < 0.05$ ). Figure 1 displays the linear relationship with an increase in timing to reoperation to complication rate.

## DISCUSSION

Management of CM1 has been controversial, with various purported surgical techniques, each with advantages and disadvantages.<sup>[12-17]</sup> Studies have identified risk factors, such as older age, male gender, and hydrocephalus, as contributors to increased complications; however, to the best of our



**Figure 1: Complication rate as a result of delayed procedure 1 = same day, 2 = 1-day delay, 3 = 2-day delay, 4 = 3-day delay, 5 = 4-7-day delay, 6 = 8-14-day delay, 7 = 14+ day delay**

knowledge, there have been none which evaluated time to intervention. This represents a potentially crucial component in a complex treatment paradigm that may shift management altogether for this patient population. The results of our study demonstrated that patients who were operated on the SD had a significantly lower incidence of complications, length of hospital stay, and total hospital charges when compared to patients who had delayed procedures.

There have been various studies that have evaluated additional risk factors and outcomes for patients with CM1.<sup>[21,22]</sup> A study by Bhimani *et al.* reviewed an international database of 672 patients and found that obesity, male gender, and the American Society of Anesthesiologists class were predictive of reoperations as well as readmissions.<sup>[21]</sup> Furthermore, the authors found that the CSF leak was the most common reason for readmissions. These factors have been demonstrated across various surgical subspecialties to negatively impact operative outcomes, many of which may not effectively be modified in the short term on hospital admission. Therefore, it is possible that early intervention may be effective at reducing overall complications while controlling for certain modifiable risk factors to optimize surgery.

While we were unable to find any studies that have specifically reviewed time to procedure to determine outcomes, a study by Vedantam *et al.* evaluated 1459 patients using the National Surgical Quality Improvement Program database to identify the factors that were associated with adverse perioperative outcomes.<sup>[23]</sup> The authors found that on multivariate analysis, hospital stay >5 days was an independent risk factor associated with adverse perioperative events. This study supports our

**Table 2: Postoperative complications by timing to operation**

| Timing to operation | Postoperative complications |        |
|---------------------|-----------------------------|--------|
|                     | Odds ratio                  | P      |
| Same day            | 0.88 (0.7-0.9)              | 0.038  |
| 1-day delay         | 1.2 (1.0-1.5)               | 0.009  |
| 2-day delay         | 1.3 (1.0-1.7)               | 0.039  |
| 3-day delay         | 1.4 (1.04-2.0)              | 0.025  |
| 4-7-day delay       | 2.3 (1.8-3.0)               | <0.001 |
| 8-14-day delay      | 3.1 (2.3-4.2)               | <0.001 |
| 15+ day delay       | 4.7 (3.4-6.6)               | <0.001 |

findings that delays to surgery for this patient population may be a risk factor for increased peri- and postoperative complications.

There were some limitations to our study, mostly the retrospective nature of our review. In addition, the use of national databases, such as KID, imparts some inherent limitations which that may impact some of the results and findings.<sup>[24]</sup> One such limitation is that this database only includes inpatient complications and events, which may result in an underrepresentation of true long-term complication rates. Furthermore, reliance on the accuracy of diagnosis and procedure coding may lead to inaccuracies as well as over- or under-estimations of potential associations. However, even with these limitations, a large database allows us to have an adequately powered study which enhances the reliability and applicability of our results.

With the true prevalence of hindbrain abnormalities underestimated due to varied presentation, the development of enhanced diagnostics may result in the identification of additional patients with CM1 requiring surgical management. It has been reported that at 90 days postoperatively, patients who experienced medical or surgical complications following the initial procedure incurred markedly higher hospital costs, by >2-fold.<sup>[9,25]</sup> These complications result in marked risks to the patient with potentially increased morbidity and mortality, as well as represented tremendous financial burden to hospital systems. Our study was able to identify timing to intervention from admission within the range of 8–14 days as an independent risk factor for increased complications and hospital expenditures postoperatively. These findings may be utilized to provide surgeons with information regarding optimal timing for the surgical management of CM1 in an attempt to reduce complication rates, while optimizing additional patient risk factors. Further prospective randomized trials are necessary to be able to validate the results of our findings.

## CONCLUSIONS

Patients with CM1 operated on the SD had lower LOS and total hospital charges than all delay groups. These

patients were also significantly less likely to develop sepsis, respiratory complications, and anemia postoperatively. Delaying procedure from admission within the range of 8–14 days was associated with greater overall complications as well as greater nervous system complications.

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## Conflicts of interest

There are no conflicts of interest.

## REFERENCES

- Grahovac G, Pundy T, Tomita T. Chiari type I malformation of infants and toddlers. *Childs Nerv Syst* 2018;34:1169-76.
- Hidalgo JA, Tork CA, Varacallo M. Arnold Chiari Malformation. Treasure Island (FL): StatPearls; 2020.
- Arnautovic A, Splavski B, Boop FA, Arnautovic KI. Pediatric and adult Chiari malformation Type I surgical series 1965-2013: A review of demographics, operative treatment, and outcomes. *J Neurosurg Pediatr* 2015;15:161-77.
- 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.
- Dyste GN, Menezes AH, van Gilder JC. Symptomatic Chiari malformations. An analysis of presentation, management, and long-term outcome. *J Neurosurg* 1989;71:159-68.
- Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Speer W. Chiari I malformation redefined: Clinical and radiographic findings for 364 symptomatic patients. *Neurosurgery* 1999;44:1005-17.
- Hidalgo ET, Dastagirzada Y, Orillac C, Kvint S, North E, Bledera R, et al. Time to resolution of symptoms after suboccipital decompression with duraplasty in children with Chiari malformation type I. *World Neurosurg* 2018;117:e544-51.
- Peach B. The arnold-chiari malformation; morphogenesis. *Arch Neurol* 1965;12:527-35.
- Greenberg JK, Ladner TR, Olsen MA, Shannon CN, Liu J, Yarbrough CK, et al. Complications and resource use associated with surgery for chiari malformation type 1 in adults: A population perspective. *Neurosurgery* 2015;77:261-8.
- 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.
- Poretti A, Ashmawy R, Garzon-Muvdi T, Jallo GI, Huisman TA, Raybaud C. Chiari type I deformity in children: Pathogenetic, clinical, neuroimaging, and management aspects. *Neuropediatrics* 2016;47:293-307.
- Alexander H, Tsering D, Myseros JS, Magge SN, Oluigbo C, Sanchez CE, et al. Management of Chiari I malformations: A paradigm in evolution. *Childs Nerv Syst* 2019;35:1809-26.
- El-Ghandour NM. Long-term outcome of surgical management of adult Chiari I malformation. *Neurosurg Rev* 2012;35:537-46.
- Giammattei L, Messerer M, Daniel RT, Aghakhani N, Parker F. Long term outcome of surgical treatment of Chiari Malformation without syringomyelia. [published online ahead of print, 2017 Jul 4]. *J Neurosurg Sci*. 2017;10.23736/S0390-5616.17.04063-2.
- Entezami P, Gooch MR, Poggi J, Perloff E, Dupin M, Adamo MA. Current management of pediatric chiari type 1 malformations. *Clin Neurol Neurosurg* 2019;176:122-6.
- Leon TJ, Kuhn EN, Arynchyna AA, Smith B, Tubbs RS, Johnston JM,

- et al.* Patients with “benign” Chiari I malformations require surgical decompression at a low rate. *J Neurosurg Pediatr* 2019;23:498-506.
17. Giammattei L, Borsotti F, Parker F, Messerer M. Chiari I malformation: Surgical technique, indications and limits. *Acta Neurochir (Wien)* 2018;160:213-7.
  18. 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;110:460-740.
  19. Strahle J, Muraszko KM, Kapurch J, Bapuraj JR, Garton HJ, Maher CO. Natural history of Chiari malformation Type I following decision for conservative treatment. *J Neurosurg Pediatr* 2011;8:214-21.
  20. Aliaga L, Hekman KE, Yassari R, Straus D, Luther G, Chen J, *et al.* A novel scoring system for assessing Chiari malformation type I treatment outcomes. *Neurosurgery* 2012;70:656-64.
  21. Bhimani AD, Esfahani DR, Denyer S, Chiu RG, Rosenberg D, Barks AL, *et al.* Adult chiari I malformations: An analysis of surgical risk factors and complications using an international database. *World Neurosurg* 2018;115:e490-500.
  22. Hekman KE, Aliaga L, Straus D, Luther A, Chen J, Sampat A, *et al.* Positive and negative predictors for good outcome after decompressive surgery for Chiari malformation type 1 as scored on the Chicago Chiari Outcome Scale. *Neurol Res* 2012;34:694-700.
  23. Vedantam A, Mayer RR, Staggers KA, Harris DA, Pan IW, Lam SK. Thirty-day outcomes for posterior fossa decompression in children with Chiari type 1 malformation from the US NSQIP-Pediatric database. *Childs Nerv Syst* 2016;32:2165-71.
  24. Murphy M, Alavi K, Maykel J. Working with existing databases. *Clin Colon Rectal Surg* 2013;26:5-11.
  25. Lam SK, Mayer RR, Luerssen TG, Pan IW. Hospitalization cost model of pediatric surgical treatment of chiari type 1 malformation. *J Pediatr* 2016;179:204-100.