Original Study

Developments in the treatment of Chiari type 1 malformations over the past decade

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Background: Chiari malformations type 1 (CM-1), a developmental anomaly of the posterior fossa, usually presents in adolescence or early adulthood. There are few studies on the national incidence of CM-1, taking into account outcomes based on concurrent diagnoses. To quantify trends in treatment and associated diagnoses, as retrospective review of the Kid's Inpatient Database (KID) from 2003-2012 was conducted.

Methods: Patients aged 0–20 with primary diagnosis of CM-1 in the KID database were identified. Demographics and concurrent diagnoses were analyzed using chi-squared and *t*-tests for categorical and numerical variables, respectively. Trends in diagnosis, treatments, and outcomes were analyzed using analysis of variance (ANOVA).

Results: Five thousand four hundred and thirty-eight patients were identified in the KID database with a primary diagnosis of CM-1 (10.5 years, 55% female). CM-1 primary diagnoses have increased over time (45 to 96 per 100,000). CM-1 patients had the following concurrent diagnoses: 23.8% syringomyelia/syringobulbia, 11.5% scoliosis, 5.9% hydrocephalus, 2.2% tethered cord syndrome. Eighty-three point four percent of CM-1 patients underwent surgical treatment, and rate of surgical treatment for CM-1 increased from 2003–2012 (66% to 72%, P<0.001) though complication rate decreased (7% to 3%, P<0.001) and mortality rates remained constant. Seventy percent of surgeries involved decompression-only, which increased neurologic complications compared to fusions (P=0.039). Cranial decompressions decreased from 2003–2012 (42.2–30.5%) while spinal decompressions increased (73.1–77.4%). Fusion rates have increased over time (0.45% to 1.8%) and are associated with higher complications than decompression-only (11.9% *vs.* 4.7%). Seven point four percent of patients experienced at least one peri-operative complication (nervous system, dysphagia, respiratory most common). Patients with concurrent hydrocephalus had increased; nervous system, respiratory and urinary complications (P<0.006) and syringomyelia increased the rate of respiratory complications (P=0.037).

Conclusions: CM-1 diagnoses have increased in the last decade. Despite the decrease in overall complication rates, fusions are becoming more common and are associated with higher peri-operative complication rates. Commonly associated diagnoses including syringomyelia and hydrocephalus, can dramatically increase complication rates.

Keywords: Chiari; Chiari malformations type 1 (CM-1); complication rates; spinal decompression; pediatrics

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Introduction

Named after Austrian pathologist Hans Chiari, the Chiari malformations (CM) are a variety of structural deformities of the lower skull and cerebellum that range in severity and incidence (1). The most common and least serious variation of the Chiari syndrome is type 1. Chiari malformations type 1 (CM-1) patients aged 15 years and younger experience a 5 mm or greater displacement of the cerebellar tonsils below the foramen magnum, while patients over 15 years of age experience a 6 mm or greater displacement (2-5). These displacements typically give rise to brainstem and spinal cord compression and disrupted flow of cerebrospinal fluid (CSF) and are also thought to lead to reduced volume of the posterior fossa (3,6,7). The etiology of CM remains uncertain; however, several theories have been put forth. For instance, it has been suggested that interrupted cranial development, especially in the hindbrain, results in these structural deformities (2,4,8,9). Another theory supposes that irregular circulation of CSF alters intracranial pressure and causes CM (2,10). The pressure on the brainstem and cerebellum, caused by fetal hydrocephalus, has also been suggested as a potential cause (4). There is no consensus regarding the pathogenesis of the Chiari syndrome, as its etiology is still being researched.

While it is known that CM-1 is the most common variety of CM, the incidence of this medical condition is still relatively difficult to estimate. Studies on this matter typically report incidences of CM-1 that range from 0.56–1.00% (3,5,6,11,12), making CM-1 a fairly uncommon condition. Symptoms typically do not present until the third decade of life (12), and headaches are the most commonly-reported symptom amongst symptomatic CM-1 patients (2,6,11). However, in many cases, CM-1 patients experience no symptoms at all and find any sort of therapeutic intervention unnecessary (12).

For those CM-1 patients who do experience symptoms, there are both surgical and non-surgical treatment options. Initially, many patients undergo physical therapy or therapeutic injections to ease the burden of headaches (5). However, if symptoms are severe or persistent, there are treatment paths that include surgical intervention. The most common surgical treatment for CM-1 patients is a posterior fossa decompression surgery with or without duraplasty (2,4,13). There is currently no consensus on whether or not a duraplasty should be included in the treatment of CM-1 (8,11), although one recent study showed that the inclusion of a duraplasty is inconsequential and only raises the chances of complication (4). In some cases, however, different surgeries that target the perceived cause of CM-1 may be preferred (2). For instance, surgeons who believe disrupted CSF dynamics are the primary cause of CM-1 may seek to restore CSF flow through different types of shunt procedures (2,4,14).

To date, completed studies on the Chiari syndrome have attempted to recommend an optimal treatment path for patients by studying the efficacy of various surgeries. These studies, however, primarily consider short-term postoperative conditions and fail to examine the preoperative and intraoperative conditions of CM-1 patients. Long-term clinical outcomes of CM-1 patients are also underreported and subsequently understudied (11), and an evaluation of surgical CM-1 patients reveals that rates of symptom reoccurrence are as high as 35% (13). The prevalence of symptom reoccurrence, paired with understudied aspects of CM-1 cases, illustrates a clear need for more expansive research on the intricacies of CM-1. The present study evaluates over 5,000 patients on the larger, perioperative scale, which not only enhances our understanding of longer-term trends of CM-1 cases, but also begins to fill the scientific community's need for more extensive and larger-scale analyses of CM-1 patients.

Therefore, the aim of this study is to quantify trends in CM-1 diagnoses and treatments using the paediatric-specific Kids' Inpatient Database (KID) to assess fluctuations in diagnoses, treatment options, and outcomes of patients with CM-1.

Methods

Data source

The Kids' Inpatient Database (KID) is the United States' most expansive publicly-available all-payer pediatric (age <21 at admission) inpatient medical database. Created by the Agency for Healthcare Research and Quality's Healthcare Cost and Utilization Project (HCUP), the KID includes information regarding complicated and uncomplicated births, as well as other inpatient procedures from community, non-rehabilitation hospitals. The KID database contains 107 data elements, using *International Classification of Disease*, *Ninth Revision*, *Clinical Modification* (ICD-9-CM) format to code all diagnoses and procedures until 2015. With over 3 million hospital stays per 3-year database, the KID facilitates analyses of diagnostic and therapeutic incidences using HCUP-provided trend weights (15,16). A detailed synopsis of the KID can be found at

Table 1 Surgical CM-1 patient demographics by spinal decompression cohort

Demographics	Spinal decompression (N=3,596)	Non-spinal decompression (N=941)	P value
Age, years	10.47±5.65	10.62±5.53	0.455
CCI	0.21±0.49	0.18±0.49	0.174
Gender (female), n (%)	1,980 (55.1)	521 (55.4)	0.443
LOS, days	4.01±4.89	4.10±3.06	0.583
Mortality, n (%)	5 (0.1)	0	0.313

CM-1, Chiari malformations type 1; CCI, Charlson comorbidity index; LOS, length of stay.

(https://www.hcup-us.ahrq.gov/kidoverview.jsp).

Patient sample

Patients in the KID database with a diagnosis of CM-1 were found using the ICD-9 code 348.4. A sub-analysis compared CM-1 malformation patients who had undergone a spinal decompression procedure (ICD-9 code 03.09) to CM-1 patients without spinal decompression performed.

Statistical analysis

IBM SPSS Statistics version 23.0 (IBM Corp., Armonk, NY, USA) was used to perform all descriptive and comparative statistics. We quantified trends of CM-1 treatments and concurrent diagnoses in the United States. Trends in demographics, procedures, and complications were analyzed using analysis of variance (ANOVA). Differences between CM-1 patients who underwent spinal decompression surgery and CM-1 patients who had not were assessed using Chi-squared and *t*-tests that quantified correlations among demographics, CM-1 treatments, concurrent diagnoses, complications, and secondary diagnoses. P values that were less than 0.05 were considered statistically significant.

Results

CM-1 patient demographics and comorbidities

Five thousand four hundred and thirty-eight CM-1 patients were identified in the KID from 2003–2012. The prevalence of CM-1 diagnoses has increased from 2003 to 2012, from 45 to 96 per 100,000 (P<0.001). Fifty-four point six percent of these patients were female, and the average age of these CM-1 patients was 10.51±5.71 years. Overall, 10.9% patients accounted for infantile CM-1 cases, while 36.1% of patients and 47.0% of patients represented juvenile and adolescent CM-1 cases, respectively. Fifty-nine percent of all CM-1 primary patients were white. The average length of stay (LOS) for all CM-1 patients was 4.02±7.78 days, and the average Charlson comorbidity index (CCI) for all patients was 0.213±0.519. The breakdown of demographic results based on CM-1 patients who underwent spinal decompressions and those who did not can be found in *Table 1*.

As a reference, the KID database as a whole contains 12,718,381 patients with an average age of 6.88 years (range, 0–20 years) and is 53.8% female. The most common diagnoses in the entire KID database are pneumonia, respiratory syncytial virus-induced bronchitis, asthma, and appendicitis. One point seven percent of patients in the KID database have a diagnosis of depression and 0.8% have a diagnosis of anxiety.

From 2003–2012, the average age of all CM-1 patients steadily increased (2003: 10.02±5.57 *vs.* 2012: 10.76±5.71, P=0.004) (*Table 2*). The average CCI of all surgical patients also increased over time, from 0.16±0.46 in 2003 to 0.25±0.57 in 2012 (P<0.001) (*Table 2*). Over time, CM-1 patients, experienced a shorter average LOS, as the average LOS in 2003 was 4.12±2.84 days and in 2012 was 3.80±4.22 days (P=0.016) (*Table 2*).

Concurrent diagnoses in CM-1 patients

Overall, 53.9% of all CM-1 patients were diagnosed with a concurrent condition. The most common concurrent diagnoses of all CM-1 patients were syringomyelia (23.8%), scoliosis (11.5%), hydrocephalus (5.9%), and various functional digestive disorders (5.5%). The most common secondary diagnoses were syringomyelia/syringobulbia (18.7%), asthma (3.4%), obstructive hydrocephalus (3.3%), and idiopathic scoliosis (2.9%) (*Table 3*). Sixty-three point seven percent of patients who underwent spinal decompressions were diagnosed with a concurrent

Table 2 CM-1 patient demographics over time

Demographics	2003 (N=886)	2006 (N=1,133)	2009 (N=1,478)	2012 (N=1,941)	P value
Age, years	10.02±5.57	10.27±5.68	10.65±5.81	10.76±5.71	0.004*
In-hospital mortality, n (%)	9 (1.0)	2 (0.2)	12 (0.8)	10 (0.5)	0.065
CCI	0.16±0.46	0.21±0.50	0.20±0.48	0.25±0.57	<0.001*
Length of stay, days	4.12±2.84	4.36±5.75	3.99±5.52	3.80±4.22	0.016*

*, significance of P<0.05. CM-1, Chiari malformations type 1; CCI, Charlson comorbidity index.

Table 3 Concurrent diagnoses for CM-1 patients, spinal decompression cohort, and non-spinal decompression cohort

Concurrent diagnosis	CM-1 patients (N=5,438)	Spinal decompression (N=3,596)	Non-spinal decompression (N=941)	P value
Depression, n (%)	117 (2.2)	76 (2.2)	41 (4.4)	0.768
Anxiety, n (%)	100 (1.8)	69 (1.9)	31 (3.3)	0.595
Scoliosis, n (%)	628 (11.5)	457 (12.7)	171 (18.2)	0.024*
Klippel-Feil, n (%)	17 (0.3)	11 (0.31)	6 (0.64)	0.901
Spina bifida, n (%)	11 (0.2)	9 (0.25)	2 (0.21)	0.353
Torticollis, n (%)	28 (0.5)	19 (5.3)	9 (0.96)	0.846
Functional digestive disorders, n (%)	301 (5.5)	209 (5.8)	92 (9.8)	0.234
Tethered cord, n (%)	118 (2.2)	85 (2.4)	33 (3.5)	0.201
Hydrocephalus, n (%)	319 (5.9)	189 (5.3)	130 (13.8)	0.009*
Syringomyelia, n (%)	1,295 (23.8)	991 (27.6)	304 (32.3)	<0.001*

*, significance of P<0.05. CM-1, Chiari malformations type 1.

condition, while only 44.46% of patients who did not have a spinal decompression had concurrent diagnoses. The incidences of most concurrent conditions were largely the same for the two groups (*Table 3*). However, both scoliosis (12.7% *vs.* 18.2%, P=0.024) and syringomyelia (27.6% *vs.* 32.3%, P<0.001) were more common in patients who underwent spinal decompression surgeries (*Table 3*).

CM-1 surgical treatment

Overall, 83.4% of all CM-1 patients in the KID database underwent surgical procedures (*Table 4*). CM-1 surgical treatments included the following procedures: cranial decompressions, craniectomies, meninges repairs, cranial nerve decompression, spinal decompressions, fusion surgeries, other brain lesion excisions, and other spinal cord operations (*Table 4*). Patients were grouped on the basis of undergoing a spinal decompression: 3,569 underwent a spinal decompression and 941 underwent other surgical procedures (*Table 4*). One hundred percent of patients who did not undergo spinal decompressions underwent cranial decompressions, while 13% of patients who had undergone spinal decompressions underwent cranial decompressions (P<0.001) (*Table 4*).

The percentage of all CM-1 patients who underwent any sort of surgery relating to their primary conditions fluctuated over time, but ultimately increased by approximately 4% (2003: 81.3% *vs.* 2012: 85.0%, P=0.074) (*Table 5*). There were notable changes in the rates of simple fusions, cranial decompressions, and spinal decompressions/laminectomies. Simple fusion rates (2003: 0.2% *vs.* 2012: 1.5%, P=0.021) and spinal decompression/laminectomy rates (2003: 60.8% *vs.* 2012: 69.0%, P<0.001) increased over time, while cranial decompression rates decreased over time (2003: 33.6% *vs.* 2012: 23.0%, P<0.001) (*Table 5*). Craniectomy rates fluctuated over the nine-year span, but the 2012 rate was ultimately similar to the 2003 rate (2003: 11.6% *vs.* 2006: 9.5% *vs.* 2009: 14.9% *vs.* 2012: 11.1%, P<0.001) (*Table 5*).

Complications of CM-1 patients

Table 4 Surgical procedures of CM-1 patients, spinal decompression cohort, and non-spinal decompression cohort

Surgical procedure	CM-1 patients (N=5,438)	Spinal decompression (N=3,596)	Non-spinal decompression (N=941)	P value
Cranial decompression, n (%)	1,410 (25.9)	469 (13.0)	941 (100.0)	<0.001*
Craniectomy, n (%)	647 (11.9)	467 (13.0)	31 (3.3)	<0.001*
Meninges repair, n (%)	2,734 (50.3)	2,169 (60.3)	309 (32.8)	<0.001*
Other brain lesion excision, n (%)	249 (4.6)	144 (4.0)	32 (3.4)	0.226
Other spinal cord operations, n (%)	385 (7.1)	295 (8.2)	49 (5.2)	0.001*
Cranial nerve decompression, n (%)	129 (2.4)	32 (0.9)	97 (10.3)	<0.001*
Fusion surgery, n (%)	36 (0.7)	38 (1.1)	12 (1.3)	<0.001*

*, significance of P<0.05. CM-1, Chiari malformations type 1.

Table 5 CM-1 patient procedures over time

Surgical procedure	2003 (N=886)	2006 (N=1,133)	2009 (N=1,478)	2012 (N=1,941)	P value
Any surgery, n (%)	720 (81.3)	943 (83.2)	1,224 (82.8)	1,650 (85.0)	0.074
Anterior fusion, n (%)	10 (1.1)	21 (1.9)	17 (1.2)	26 (1.3)	0.408
Posterior fusion, n (%)	2 (0.2)	2 (0.2)	3 (0.2)	4 (0.2)	0.996
Simple fusion, n (%)	2 (0.2)	18 (1.6)	20 (1.4)	30 (1.5)	0.021*
Complex fusion, n (%)	0	5 (0.4)	2 (0.1)	5 (0.3)	0.167
Cranial decompression, n (%)	298 (33.6)	336 (29.7)	329 (22.3)	447 (23.0)	<0.001*
Spinal decompression/laminectomy, n (%)	539 (60.8)	727 (64.2)	990 (66.9)	1,340 (69.0)	<0.001*
Craniectomy, n (%)	103 (11.6)	108 (9.5)	220 (14.9)	216 (11.1)	<0.001*
Cranial nerve decompression, n (%)	13 (1.5)	26 (2.3)	38 (2.6)	52 (2.7)	0.240
Meninges repair, n (%)	445 (50.2)	541(47.7)	732 (49.5)	1,016 (52.3)	0.088
Other brain excision, n (%)	45 (5.1)	41 (3.6)	73 (4.9)	90 (4.6)	0.341
Other spinal cord operation, n (%)	70 (7.9)	68 (6.0)	106 (7.2)	123 (6.3)	0.391

*, significance of P<0.05. CM-1, Chiari malformations type 1.

Overall, 4.8% of CM-1 patients experienced complications during their treatments for CM-1 (*Table 6*). The most common complications were nervous system complications (1.4%), dysphagia (0.8%), respiratory and digestive complications (0.5%), and acute respiratory distress syndrome (ARDS) (0.5%) (*Table 6*). The prevalence of complications was slightly higher in patients who underwent procedures other than spinal decompressions (spinal decompression: 4.7% *vs.* non-spinal decompression: 5.0%, P=0.547) (*Table 6*). Patients who did not undergo spinal decompressions experienced infection more frequently (spinal decompression: 0.1% *vs.* non-spinal decompression: 0.3%, P=0.021) (*Table 6*). The average in-hospital mortality rate across all CM-1 cases in the KID database was 0.61%, or 33 patients (*Table 2*). Patients who had previously undergone spinal decompressions experienced a higher mortality rate when compared to the non-spinal decompression group (spinal decompression: 0.8% *vs.* non-spinal decompression: 0.3%, P<0.001).

In looking at complication trends, we found that, for all CM-1 patients, complication rates decreased from 2003–2012 (2003: 5.3% *vs.* 2012: 2.6%, P<0.001) (*Table 7*). Rates of digestive complications (2003: 0.9% *vs.* 2012: 0%, P=0.001), dysphagia (2003: 1.5% *vs.* 2012: 0.2%, P<0.001), and ARDS (2003: 0.5% *vs.* 2012: 0%, P<0.001) decreased

Table 6 Complications of CM-1 Patients, spinal decompression cohort, and non-spinal decompression cohort

Complication	CM-1 patients (N=5,438)	Spinal decompression (N=3,596)	Non-spinal decompression (N=1,842)	P value
Dysphagia, n (%)	44 (0.8)	29 (0.8)	15 (0.8)	0.545
Nervous, n (%)	77 (1.4)	50 (1.4)	27 (1.5)	0.809
Cardiac, n (%)	12 (0.2)	9 (0.3)	3 (0.2)	0.761
PVD, n (%)	1 (0.1)	0	1 (0.1)	0.339
Respiratory, n (%)	29 (0.5)	19 (0.5)	10 (0.5)	0.542
Digestive, n (%)	27 (0.5)	15 (0.4)	12 (0.7)	0.307
Urinary, n (%)	13 (0.2)	8 (0.2)	5 (0.3)	0.772
Device, n (%)	3 (0.1)	2 (0.1)	1 (0.1)	0.734
Shock, n (%)	1 (0.1)	1 (0.1)	0	0.661
Hematoma/seroma, n (%)	4 (0.1)	3 (0.1)	1 (0.1)	0.583
Puncture, n (%)	14 (0.3)	10 (0.3)	4 (02)	0.784
Infection, n (%)	8 (0.1)	2 (0.1)	6 (0.3)	0.021*
Anemia, n (%)	22 (0.4)	16 (0.4)	6 (0.3)	0.654
ARDS, n (%)	29 (0.5)	18 (0.5)	11 (0.6)	0.695
Wound, n (%)	2 (0.1)	1 (0.1)	1 (0.1)	0.563
Dural tear, n (%)	21 (0.4)	17 (0.5)	4 (0.2)	0.172
PE, n (%)	1 (0.1)	1 (0.1)	0	0.661
DVT, n (%)	3 (0.1)	2 (0.1)	1 (0.1)	0.734
Any complication, n (%)	261 (4.8)	168 (4.7)	93 (5.0)	0.547

*, significance of P<0.05. CM-1, Chiari malformations type 1; PVD, peripheral vascular disease; ARDS, acute respiratory distress syndrome; PE, pulmonary embolism; DVT, deep vein thrombosis.

over time, but dural tear rates steadily increased from 2003 to 2012 (2003/2006: 0% *vs.* 2009: 0.4% *vs.* 2012: 0.7%, P=0.003) (*Table 7*). The mortality rate for all CM-1 patients also increased slightly from 2003 to 2012, but this was considered statistically insignificant (2003: 0.1% *vs.* 0.2%, P=0.582) (*Table 7*).

The overall complication rate for patients who underwent spinal decompressions decreased from 2003 to 2012 (2003: 6.9% *vs.* 2012: 2.8%, P<0.001) (*Table 8*); the overall complication rate for the non-spinal decompression cohort also decreased over time (2003: 5.6% *vs.* 2012: 3.9%, P=0.246), but this trend was considered statistically insignificant (*Table 9*). In 2009, there was a spike in the complication rates of ARDS for both cohorts: spinal decompression patients (2003: 0.7% *vs.* 2006: 0.6% *vs.* 2009: 1.0% *vs.* 2012: 0%, P=0.006) (*Table 8*) and non-spinal decompression patients (2003: 0% *vs.* 2006: 0.9% *vs.* 2009: 2.1% *vs.* 2012: 0%, P=0.019) (*Table 9*). For patients who had undergone spinal decompressions, there was a statistically significant decrease in the occurrence of digestive complications (2003: 1.1% *vs.* 2012: 0%, P=0.001) (*Table 8*); non-spinal decompression patients also saw a decreased rate of digestive complications (2003: 1.1% *vs.* 2012: 0%, P=0.081) (*Table 9*). Dysphagia rates decreased in the spinal decompression group (2003: 2.0% *vs.* 2012: 0.2%, P<0.001) (*Table 8*) and also in the non-spinal decompression cohort (2003: 1.1% *vs.* 2012: 0%, P=0.147), but the latter was considered statistically insignificant (*Table 9*). Dural tear rates for the spinal decompression cohort increased from 0% in 2003 to 0.9% in 2012 (P=0.011) (*Table 8*); dural tears also became increasingly common in the non-spinal decompression group (2003: 0% *vs.* 2012: 0.6%, P=0.496) (*Table 9*). From 2003 to 2012, the mortality rate for both groups remained nearly constant for the spinal

Table 7 Surgical CM-1 patient complications over time

Complication	2003 (N=886)	2006 (N=1,133)	2009 (N=1,478)	2012 (N=1,941)	P value
Mortality rate, n (%)	1 (0.1)	0	1 (0.1)	3 (0.2)	0.582
Dysphagia, n (%)	13 (1.5)	17 (1.5)	4 (0.3)	3 (0.2)	<0.001*
Nervous system, n (%)	11 (1.2)	13 (1.1)	18 (1.2)	22 (1.1)	0.981
Cardiac, n (%)	1 (0.1)	1 (0.1)	6 (0.4)	4 (0.2)	0.293
PVD	–	–	–	–	–
Respiratory, n (%)	8 (0.9)	6 (0.5)	6 (0.4)	5 (0.3)	0.103
Digestive, n (%)	8 (0.9)	6 (0.5)	12 (0.8)	0	0.001*
Urinary, n (%)	3 (0.3)	4 (0.4)	4 (0.3)	2 (0.1)	0.440
Device-related, n (%)	0	0	1 (0.1)	1 (0.1)	0.746
Shock, n (%)	0	0	1 (0.1)	0	0.439
Hematoma, n (%)	1 (0.1)	0	2 (0.1)	0	0.257
Puncture, n (%)	1 (0.1)	7 (0.6)	2 (0.1)	2 (0.1)	0.016*
Infection, n (%)	1 (0.1)	7 (0.6)	2 (0.1)	0	0.257
Anemia, n (%)	1 (0.1)	2 (0.2)	8 (0.5)	7 (0.4)	0.251
ARDS, n (%)	4 (0.5)	6 (0.5)	15 (1.0)	0	<0.001*
Wound, n (%)	0	0	0	1 (0.1)	0.626
Dural tear, n (%)	0	0	6 (0.4)	14 (0.7)	0.003*
PE, n (%)	0	0	0	1 (0.1)	0.626
DVT, n (%)	0	1 (0.1)	1 (0.1)	1 (0.1)	0.860
Any complication, n (%)	47 (5.3)	54 (4.8)	72 (4.9)	50 (2.6)	<0.001*

*, significance of P<0.05. CM-1, Chiari malformations type 1; PVD, peripheral vascular disease; ARDS, acute respiratory distress syndrome; PE, pulmonary embolism; DVT, deep vein thrombosis.

decompression cohort (2003: 0.2% *vs.* 2006: 0% *vs.* 2009: 0.1% *vs.* 2012: 0.2%, P=0.595) (*Table 8*) and completely constant for the non-spinal decompression cohort (2003: 0% *vs.* 2006: 0% *vs.* 2009: 0% *vs.* 2012: 0%) (*Table 9*).

Discussion

CM-1 can negatively impact quality of life and induce problematic symptomatic responses that warrant surgical intervention. Despite this, the scientific community lacks a complete understanding of how a CM-1 patient's pre-operative and intra-operative conditions correlate to post-operative outcomes. This study examined trends between patient outcomes and peri-operative conditions for 5,438 CM-1 pediatric patients using the KID. We found that the prevalence of CM-1 cases has approximately doubled from 2003–2012 (45 to 96 per 100,000, P<0.001). This increase in incidence is consistent with the recent findings from other studies (17), and researchers are speculating that these increased diagnoses could be a result of progress in magnetic resonance imaging, the primary diagnostic tool for the Chiari syndrome (3,6).

Preoperative trends

Examination of the results related to preoperative data yields interesting conclusions about the evolving demographics of primary CM-1 patients. The present study showed that not only have the average age and CCI score of surgical CM-1 patients increased over time, but the average age of CCI score of all CM-1 patients (including those not undergoing surgery) also increased from 2003

Table 8 Spinal decompression cohort complications over time

Complication	2003 (N=539)	2006 (N=727)	2009 (N=990)	2012 (N=1,340)	P value
Mortality rate, n (%)	1 (0.2)	0	1 (0.1)	3 (0.2)	0.595
Dysphagia, n (%)	11 (2.0)	13 (1.8)	2 (0.2)	3 (0.2)	<0.001*
Nervous system, n (%)	9 (1.7)	10 (1.4)	16 (1.6)	15 (1.1)	0.707
Cardiac, n (%)	0	1 (0.1)	4 (0.4)	4 (0.3)	0.426
PVD, n (%)	0	0	0	0	–
Respiratory, n (%)	6 (1.1)	6 (0.8)	4 (0.4)	3 (0.2)	0.061
Digestive, n (%)	6 (1.1)	1 (0.1)	8 (0.8)	0	0.001*
Urinary, n (%)	3 (0.6)	2 (0.3)	2 (0.2)	1 (0.1)	0.247
Device-related, n (%)	0	0	1 (0.1)	1 (0.1)	0.763
Shock, n (%), n (%)	0	0	1 (0.1)	0	0.452
Hematoma, n (%)	1 (0.2)	0	2 (0.2)	0	0.254
Puncture, n (%)	1 (0.2)	6 (0.8)	2 (0.2)	1 (0.1)	0.017*
Infection, n (%)	0	0	2 (0.2)	0	0.153
Anemia, n (%)	1 (0.2)	2 (0.3)	7 (0.7)	6 (0.4)	0.419
ARDS, n (%)	4 (0.7)	4 (0.6)	10 (1.0)	0	0.006*
Wound, n (%)	0	0	0	1 (0.1)	0.641
Dural tear, n (%)	0	0	5 (0.5)	12 (0.9)	0.011*
PE, n (%)	0	0	0	1 (0.1)	0.641
DVT, n (%)	0	0	1 (0.1)	1 (0.1)	0.763
Any complication, n (%)	37 (6.9)	39 (5.4)	54 (5.5)	38 (2.8)	<0.001*

*, significance of P<0.05. PVD, peripheral vascular disease; ARDS, acute respiratory distress syndrome; PE, pulmonary embolism; DVT, deep vein thrombosis.

to 2012. Despite this higher rate of comorbidities, CM-1 patients were staying for shorter periods of time in the hospital, which could potentially be the result of improved in-hospital care. The demographic variables measured in this study (CCI, LOS, age, race, and gender) had minimal association with whether or not patients underwent a spinal decompression. However, this study did show that patients in the spinal decompression cohort more frequently had concurrent diagnoses, especially scoliosis and syringomyelia.

Surgical trends

This study also revealed that overall surgery rates of CM-1 patients has increased by 4% from 2003 to 2012, which is a trend that mirrors the increase in CM-1 diagnoses from 2003 to 2012. These findings are consistent with those of other studies, such as the recent work of Wilkinson *et al.* (17). Based on the findings of the present study, the increased rates of fusions and spinal decompression procedures have influenced the overall surgery rate most notably despite the decreased rates of cranial decompressions.

Postoperative trends

Despite the net increase in surgeries for CM-1 patients, there was a decrease in complication rates over time for the same cohort. This finding was unexpected and could also be linked to improved in-hospital conditions. However, from 2003 to 2012, there was an overall increase in the dural tear rate. Specifically, dural tear rates were more common in patients who had undergone spinal decompressions. When patients who had undergone spinal decompressions

Table 9 Non-spinal decompression cohort complications over time

Complication	2003 (N=181)	2006 (N=216)	2009 (N=234)	2012 (N=310)	P value
Mortality rate, n (%)	0	0	0	0	–
Dysphagia, n (%)	2 (1.1)	4 (1.9)	2 (0.9)	0	0.147
Nervous system, n (%)	2 (1.1)	3 (1.4)	2 (0.9)	7 (2.3)	0.555
Cardiac, n (%)	1 (0.6)	0	2 (0.9)	0	0.251
PVD, n (%)	0	0	0	0	–
Respiratory, n (%)	2 (1.1)	0	2 (0.9)	2 (0.6)	0.536
Digestive, n (%)	2 (1.1)	5 (2.3)	4 (1.7)	0	0.081
Urinary, n (%)	0	2 (0.9)	2 (0.9)	1 (0.3)	0.509
Device-related, n (%)	0	0	0	0	–
Shock, n (%), n (%)	0	0	0	0	–
Hematoma, n (%)	0	0	0	0	–
Puncture, n (%)	0	1 (0.5)	0	1 (0.3)	0.638
Infection, n (%)	1 (0.6)	0	0	0	0.241
Anemia, n (%)	0	0	1 (0.4)	1 (0.3)	0.679
ARDS, n (%), n (%)	0	2 (0.9)	5 (2.1)	0	0.019*
Wound, n (%)	0	0	0	0	–
Dural tear, n (%)	0	0	1 (0.4)	2 (0.6)	0.496
PE, n (%)	0	0	0	0	–
DVT, n (%)	0	1 (0.5)	0	0	0.340
Any complication, n (%)	10 (5.6)	15 (6.9)	18 (7.7)	12 (3.9)	0.246

*, significance of $P<0.05$. PVD, peripheral vascular disease; ARDS, acute respiratory distress syndrome; PE, pulmonary embolism; DVT, deep vein thrombosis.

were compared more extensively to those who had not, we found that the complication rates between both cohorts were similar—with the non-spinal decompression group experiencing complications slightly more often, but that the non-spinal decompression cohort had a statistically significant higher mortality rate.

Limitations

We understand that there are several limitations in our study, all of which are primarily a result of the restrictions of the KID. For instance, the KID only includes inpatient data, which limits our access to information that could indicate long-term outcomes, such as radiographic images or other clinical data. Access to longer-term patient information may have influenced our analysis of complications that occurred after patient discharge. Furthermore, the KID does not include any sort of radiographic imaging, which potentially limits our understanding of the severity of certain cases. Similarly, the KID does not have the granularity to convey nuances of certain cases and details of procedures, such as surgical instruments used. In spite of these limitations, this study is the only of its kind that evaluates the trends of CM-1 cases on a perioperative scale from 2003 to 2012.

Conclusions

CM-1 diagnoses have increased from 2003 to 2012. This study showed that the average age and CCI of CM-1 patients entered into the KID have increased from 2003 to 2012 and that the average LOS has decreased during that time. Rates of surgical intervention of any kind for the

treatment of CM-1 have also increased during this time. However, cranial decompression rates have decreased, as the rates of fusions and spinal decompressions have increased. Complication rates have decreased overall for all primary CM-1 patients, but dural tears have become increasingly common in the KID database from 2003 to 2012.

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Footnote

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