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ORIGINAL ARTICLE

Anesthetic management of parturients with Arnold Chiari malformation-I: a multicenter retrospective study

T.R. Gruffi,[a] F.M. Peralta,[a] M.S. Thakkar,[b] A. Arif,[c] R.F. Anderson,[d] B. Orlando,[c] J.C. Coffman,[b] N. Nathan,[a] R.J. McCarthy,[e] P. Toledo,[a] A.S. Habib[d]
[a]*Department of Anesthesiology, Northwestern University Feinberg School of Medicine, Chicago, IL, United States*
[b]*Department of Anesthesiology, The Ohio State University Wexner Medical Center, Columbus, OH, United States*
[c]*Department of Anesthesiology, Icahn School of Medicine, Mount Sinai West Hospital, New York, NY, United States*
[d]*Department of Anesthesiology, Duke University Hospital, Durham, NC, United States*
[e]*Department of Anesthesiology, Rush University Medical Center, Chicago, IL, United States*

ABSTRACT
Background: Consensus regarding the safest mode of delivery and anesthetic management for parturients with Arnold Chiari malformation-I (ACM-I) remains controversial. This study assessed their anesthetic management and reported anesthetic complications during hospitalization for delivery.
Methods: This was a multicenter, retrospective, cohort study of patients with ACM-I undergoing vaginal or cesarean delivery. Data were obtained from the electronic databases of four United States academic institutions using International Classification of Diseases (ICD) codes from 2007–2017 at three sites and 2004–2017 at one site. The primary outcome was anesthetic complications.
Results: Data were analyzed for 185 deliveries in 148 patients. Diagnosis of ACM-I was made prior to delivery in 147 (80%) cases. Pre-delivery neurosurgical consultation for management of ACM-I was performed in 53 (36%) patients. Pre-existing symptoms were recorded for 89 (48%) of the deliveries. Vaginal deliveries occurred in 80 (43%) cases, and 62 women (78%) received neuraxial labor analgesia. Cesarean delivery was performed in 105 (57%) cases, of which 70 women (67%) had neuraxial anesthesia and 34 (32%) received general anesthesia. Post-dural puncture headache was reported in three (2%) patients who had neuraxial anesthesia, and in two (12%) patients with syringomyelia. There was one (3%) reported case of aspiration pneumonia with general anesthesia.
Conclusions: The findings suggest that anesthetic complications occur infrequently in patients with ACM-I regardless of the anesthetic management. Although institutional preference in anesthetic and obstetric care appears to drive patient management, the findings suggest that an individualized approach has favorable outcomes in this population.

Keywords: Anesthesia; Obstetric; Neuraxial; General anesthesia; Arnold Chiari malformation; Multicenter; Retrospective

Introduction

Arnold Chiari malformations (ACM) are congenital anomalies in which the cerebellum herniates through the foramen magnum, displacing the lower pons and medulla.[1] Of the four types described, type I (ACM-I) is the most common with a prevalence of up to 0.7% of the general population.[2] The condition is associated with an impaired flow of cerebrospinal fluid (CSF) from the fourth ventricle and dynamic or static herniation of brain tissue.[2] The pressure gradient generated can cause an abnormal cavitation within the spinal cord known as syringomyelia, which can be present in 25% of patients (Fig. 1).[3] Parturients with type I are usually not diagnosed until adulthood and may be asymptomatic or may manifest symptoms, including headache, ataxia, and sensorimotor impairments of the extremities.[4]

Anesthetic and obstetric management of parturients with ACM-I remains controversial. Hopkins et al. showed that CSF pressure increased with uterine contractions by a mean of 2.5 mmHg,[5] and Marx et al. demonstrated that the elevation of intracranial pressure (ICP) is much greater during the second stage of labor.[6] From a neurologic standpoint, obstetricians can be guided by neurosurgeons and neurologists about the safest mode of delivery, which may be especially prudent if a patient has symptoms of increased ICP.[4]

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Correspondence to: F.M. Peralta. Department of Anesthesiology, Northwestern University Feinberg School of Medicine, 251 E Huron St, Suite F5-704, Chicago, Illinois, 60611 USA.
E-mail address: feyce.peralta@northwestern.edu

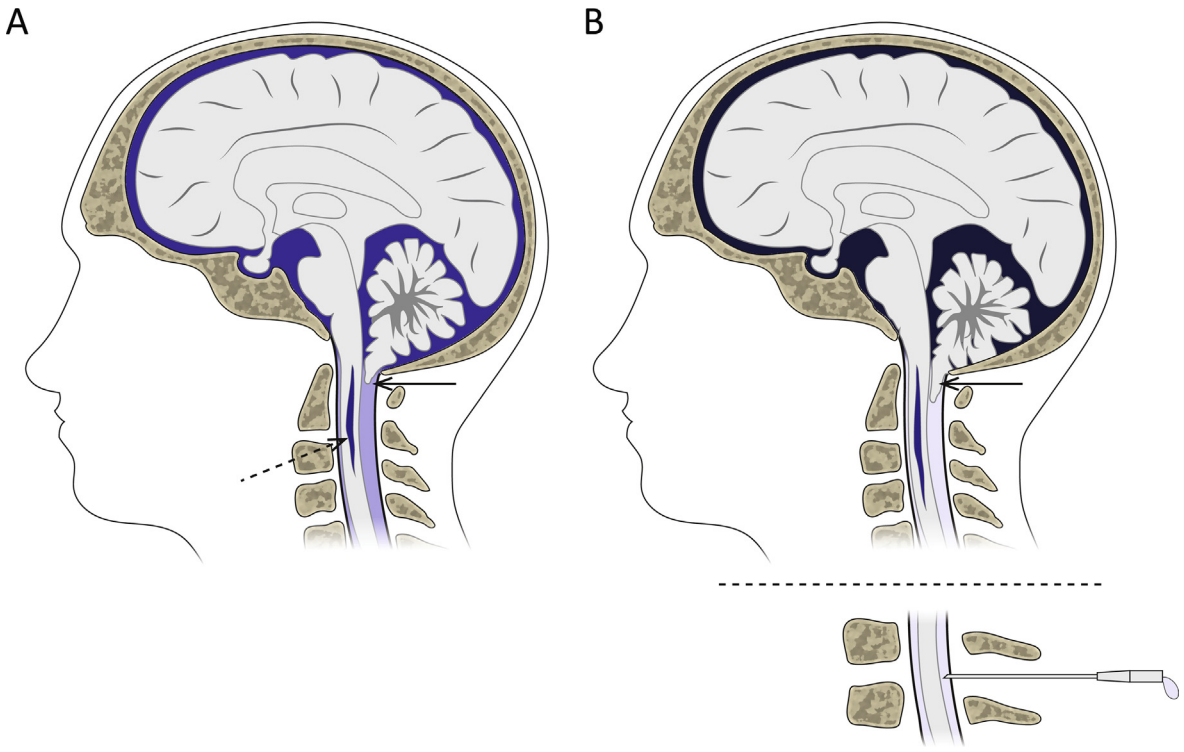

Fig. 1 Schematic representation of Arnold Chiari malformation-I. In panel A: the dotted arrow referring to the syrinx, the solid arrow referring to the Chiari malformation with herniation of the cerebellum into the foramen magnum. Despite impaired cerebrospinal fluid flow into the cervical spinal canal, there is not an excessive pressure gradient between the brain and spinal cord. In panel B: When cerebrospinal fluid pressure drops due to a dural tear (depicted with the needle) the cerebellar tonsil extends into the foramen magnum blocking the flow of cerebrospinal fluid into the cervical spinal canal (solid arrow), increasing the cerebrospinal fluid pressure exerted on the brain. In addition the decrease in pressure in the spinal canal causes the fluid pressure in the syrinx to increase, expanding the size of the syrinx in the spinal cord

While there are several advantages of using neuraxial techniques in the parturient, anesthesiologists must determine if they are safe in this particular group of patients. An unintentional dural puncture or perhaps even an uncomplicated spinal anesthetic might produce a CSF pressure gradient between brain and spinal cord with resultant cerebral herniation.[7–9] General anesthesia with laryngoscopy and tracheal intubation may also increase ICP or the CSF pressure gradient.[4]

Since no prospective randomized trial is likely to be conducted for practical reasons, the majority of the literature regarding anesthetic management of parturients with ACM-I is case reports and smaller case series,[10–19] literature reviews,[3,19,20] and algorithms.[4,21] The purpose of this study was to conduct a larger review of the anesthetic management and related complications of parturients with ACM-I who had undergone vaginal or cesarean delivery at four academic medical centers in the United States (US).

Methods

We conducted an institutional review board-approved, multicenter, retrospective, cohort study to evaluate anesthetic practices for labor analgesia and anesthesia in patients with ACM-I undergoing vaginal or cesarean delivery. Data were obtained using International Classification of Diseases (ICD)-9 codes 741.0 and 348.4, and ICD-10 code Q07.0 from the electronic databases of four US academic institutions: Northwestern University, The Ohio State University, Mount Sinai Health System, and Duke University. The electronic search was supplemented by manual chart review as needed. Implementation dates of electronic medical records differed in the institutions, and so the study period was January 1st 2007 to June 1st 2017 at three sites, and March 1st 2004 to June 1st 2017 at one site.

The inclusion criterion was any pregnant patient with a diagnosis of ACM-I who delivered at the centers during the study period. There were no exclusion criteria. Data collected included: maternal demographics; ACM symptoms, time of diagnosis (before or after delivery), surgical intervention prior to delivery; obstetric management and route of delivery; anesthetic management and anesthetic complications during hospitalization. Data were analyzed by delivery rather than by patient: each delivery was regarded and analyzed as a separate entity.

Table 1 Patient characteristics

Age in years (range)	30 (27–33)
Race/ethnicity[a]	
White Non-Hispanic	97 (66)
Black Non-Hispanic	18 (12)
Other Hispanic	12 (8)
White Hispanic	9 (6)
Other Non-Hispanic	5 (3)
Declined/Unknown	3 (2)
Multiracial Non-Hispanic	2 (1)
Asian Non-Hispanic	1 (1)
Multiracial Hispanic	1 (1)
Parity[a]	
0	72 (39)
1	67 (36)
≥2	46 (25)
Gestational age in weeks + days (range)[b]	38+6 (37+3–39+5)
ACM diagnosis before delivery[b]	147 (80)
Neurosurgical intervention before delivery[b,c]	53 (36)
Pre-existing symptoms[b]	89 (48)

ACM: Arnold Chiari malformation.
[a]n (%) calculated by patient number.
[b]n (%) calculated by delivery number.
[c]Cervical decompression or shunt placement.

The primary outcome was anesthetic complications during hospitalization for delivery. Descriptive statistics were performed.

Results

Data were collected and analyzed for 185 deliveries in 148 patients. Patient characteristics are shown in Table 1. Body mass index (BMI) was recorded for 167 of 185 deliveries. The average recorded BMI was 33.2 kg/m^2, with a range from 20.3 to 62.7 kg/m^2.

Anesthetic and obstetric data in Table 2 are divided into labor description, mode of delivery, and type of analgesia or anesthesia. Vaginal delivery occurred in 80 (43%) and cesarean delivery in 105 (57%) of cases. Indications for cesarean delivery are shown in Table 3. Repeat cesarean delivery was the most frequent reason, followed by neurosurgical or neurological recommendation. The anesthetic techniques used for each delivery type are shown in Table 4. Of note, neuraxial procedures were performed for 135 (73%) deliveries. There were 67 (50%) epidural catheters, 39 (29%) single-shot spinal anesthetics, and 29 (21%) combined spinal-epidural (CSE) catheters. Three of these epidural catheters were converted to general anesthesia for cesarean delivery, making the total number of general anesthetics 34 (18%). A sub-analysis was performed on patients with ACM-I and syringomyelia. The mode of delivery and type of anesthetic in each of these 17 deliveries is shown in Table 5.

There were no reported catastrophic neurological complications. Of the 135 patients, three (2.2%, 95% CI 0.5 to 6.4%) had post-dural puncture headache

Table 3 Indications for cesarean delivery of parturients with Arnold Chiari malformation

Indication for cesarean delivery	Number of deliveries (%)
Repeat cesarean delivery	36 (35)
Neurosurgery/Neurology recommendation	22 (21)
Failure to progress	13 (12)
Non-reassuring fetal heart rate	12 (12)
Malpresentation	9 (8)
Not documented	7 (7)
Macrosomia	2 (2)
Abnormal placentation	2 (2)
Patient request	1 (1)

Table 2 Anesthetic and obstetric practice patterns among four academic medical institutions

Institution [number of deliveries]	A [65]	B [27]	C [62]	D [31]
Labor description				
Spontaneous	35 (54)	2 (7)	17 (27)	13 (42)
Induction of labor	14 (21)	3 (11)	17 (27)	9 (29)
Elective cesarean	16 (25)	22 (81)	28 (46)	9 (29)
Mode of delivery				
Vaginal – spontaneous	32 (49)	5 (18)	26 (42)	18 (58)
Vaginal – operative	5 (8)	0	0	0
Cesarean	28 (43)	22 (82)	36 (58)	13 (42)
Analgesic/anesthetic technique				
Neuraxial	56 (86)	15 (55)	38 (61)	24 (77)
General	6 (9)	11 (41)	15 (24)	2 (6.5)
IV	2 (3)	1 (4)	4 (7)	2 (6.5)
None	1 (2)	0 (0)	5 (8)	3 (10)

Data presented as n (%). A: Northwestern University, B: Mount Sinai Health System, C: The Ohio State University, D: Duke University. IV: intravenous.

Table 4 Combined analgesic/anesthetic choices by mode of delivery for parturients with Arnold Chiari malformation

	Anesthetic								Total
Mode of delivery	Epidural	CSE	Spinal	GA	Epidural to GA	PCA	None	Unknown	
Vaginal	48	14	0	0	0	8	9	1	80
Cesarean	16	15	39	31	3	0	0	1	105
Total	64	29	39	31	3	8	9	2	185

CSE: combined spinal-epidural; GA: general anesthesia; PCA: patient-controlled analgesia.

Table 5 Mode of delivery and anesthetic technique for the 17 deliveries of 14 patients with Arnold Chiari malformation and a syrinx

	Anesthetic				Total
Mode of delivery	Epidural	CSE	Spinal	General	
NSVD	7	2	0	0	9
Elective cesarean	0	1	2	4	7
Intrapartum cesarean	1	0	0	0	1
Total	8	3	2	4	17

NSVD: normal spontaneous vaginal delivery. CSE: combined spinal-epidural.

(PDPH) but of that same 135 patients, 17 had syringomyelia, of whom two (11.7%, 95% CI 3.8 to 43.4%) had PDPH. Of the patients with syringomyelia, one had a documented unintentional dural puncture while the other had a postpartum headache that was not reported as an unintentional dural puncture. The third patient had a single-shot spinal anesthetic. In addition, there was one (3%) reported case of aspiration pneumonia associated with general anesthesia. There were no other documented anesthetic complications.

Some charts had missing information. Eight lacked postoperative notes, 15 had no mention of pre-existing neurological symptoms, 18 had no BMI recorded, and two had no documented anesthetic techniques.

Discussion

In this large cohort study from four academic medical centers, the anesthetic management and related complications of parturients with ACM-I were analyzed. Despite the high utilization rate of neuraxial procedures, there were no reported cases of worsening neurological symptoms. The incidence of unintentional dural puncture was similar to rates reported for parturients without ACM-I, but the relative risk of PDPH may be greater in patients with syringomyelia. However, the low number of patients with documented syringomyelia and high variability in our estimates preclude definitive conclusions.[22]

Anesthetic management and complications were analyzed by number of deliveries, not by number of patients, due to possible changes in the neurological status of women who had several deliveries. Of particular note, 20% of patients did not know they had ACM-I prior to their delivery, demonstrating that mild herniation may be asymptomatic and is often an incidental finding. It is unknown if the postpartum diagnoses were made because patients became symptomatic or if the diagnosis was the result of an incidental finding.

Institutional practice differed for anesthetic technique and mode of delivery. In one hospital, most patients with ACM-I were encouraged to have a neurosurgical consultation during pregnancy, and most of these women had a cesarean delivery under general anesthesia. In the other three institutions, there was less of a trend for mode of delivery. Two institutions favored neuraxial anesthesia, whether the patient underwent vaginal or cesarean delivery.

In 2013, Choi and Tyagaraj published a review of the literature on neuraxial anesthesia techniques for parturients with ACM-I and a case report of a patient who received a CSE technique for labor analgesia without complication.[19] The review included 22 patients who did not have worsening ACM-related neurological symptoms, but it did not include patients who received general anesthesia. A case series presented by Chantigian et al. in 2002 included both neuraxial (nine patients) and general anesthetic techniques (three patients).[20] The authors found no worsening of neurological ACM-related symptoms, but one (8%) patient developed a PDPH after unintentional dural puncture, requiring an epidural blood patch. Of the nine neuraxial techniques, six (67%) were epidural catheters, one (11%) was a spinal catheter, and two (22%) were single-shot spinal anesthetics.

To our knowledge the current study is the largest evaluation to date of anesthetic management of obstetric patients with ACM-I. It further supports neuraxial techniques as a viable anesthetic option. The major limitation of our study is that it was retrospective, and a number of the electronic medical records were incomplete. Had this information been available at the time

of delivery, it might have influenced the choice of anesthetic technique and mode of delivery.

Algorithms have been created to assist in the decision process. Leffert and Schwamm produced a decision tree summarizing the critical elements for assessing the risks of neurological deterioration from neuraxial anesthesia in patients with intracranial space-occupying lesions.[4] Subsequently, in 2017, the Ghaly Obstetric Guide to Arnold-Chiari malformation Type 1 (GOGAC-1) was published to assist anesthetic decision-making for patients with varying severity of ACM-I.[20]

In conclusion, anesthetic complications occurred infrequently in patients with ACM-I regardless of the anesthetic management. An individualized approach to patient care can provide favorable outcomes in this parturient population.

Declaration of interest

The authors have no declarations to make.

Conflicts of interest

All co-authors deny any conflicts of interest.

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