Case Report

Management of parturients in active labor with Arnold Chiari malformation, tonsillar herniation, and syringomyelia

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Abstract

Background: Arnold-Chiari malformation Type 1 (ACM-1) in parturients is a topic of ongoing discussion between obstetricians and anesthesiologists. The primary unanswered question remains; How should the anesthesia provider proceed with labor analgesia and anesthesia for cesarean section when confronted with an advanced, asymptomatic, or minimally symptomatic case of ACM-1 during labor?

Case Description: A 24-year-old, ASA II, G1P0 full-term parturient presented to Labor and Delivery for vaginal delivery. A diagnosis of ACM-1 was made 12 years ago when a brain magnetic resonance imaging (MRI) was performed for right-sided numbness following a rear-end motor vehicle collision. The patient had been asymptomatic since then and had been seen by an outside neurologist frequently for the past 10 years. During the anesthesia evaluation, it was noted that she had an exaggerated patellar reflex, and a questionable left-sided Babinski; subsequently, an MRI study was requested. Review of a brain MRI demonstrated an advanced form of ACM with a 1.7 cm transtonsillar herniation and a large syrinx extending from C1 down to C5. Following a discussion with the patient, family, and primary OB team, a plan for elective cesarean section was made per neurosurgical recommendations. This was conducted uneventfully under general anesthesia. The patient had no complaints in the post-anesthesia care unit.

Conclusion: Unfamiliarity of health care providers with regards to ACM-1 parturients can be countered by increasing awareness of this condition throughout medical specialties involved in their care. The Ghaly Obstetric Guide to Arnold-Chiari malformation Type 1, along with proper training of anesthesia care providers regarding the specificities of ACM-1 parturients aids in better management and understanding of this complex condition.

Key Words: ACM-1, anesthesia, delivery, multidisciplinary, parturient

INTRODUCTION

Arnold-Chiari malformation Type 1 (ACM-1) in parturients is a topic of ongoing discussion between obstetricians and anesthesiologists. Several studies have been conducted on the optimal anesthetic management of these patients during labor and delivery, however, no consensus has been reached. "How should the anesthesia

provider proceed when confronted with an advanced, asymptomatic or minimally symptomatic case of ACM-1 during labor?," remains an unanswered clinical question.

ACM-1, the most common of all four types, is a neurosurgical condition that is characterized by downward herniation of the hindbrain into the foramen magnum.[43,44]

Arnold-Chiari malformation (ACM) was first identified in 1883 by Cleland, and was first decribed and classified into four types in 1891 by Chiari. The mechanism of ACM-1 remains uncertain; however, studies support the role of the posterior cranial fossa morphometry, describing underdevelopment of the occipital bone, which causes overcrowding of the normally developed hindbrain in the posterior cranial fossa.[1,8,32] Impaired cerebrospinal fluid (CSF) flow is a common feature among all four types of ACM. It is believed that foramen magnum abnormality causes intermittent obstruction of the CSF outflow from the fourth ventricle. The consequent development of a pressure gradient, with higher pressures in the cranium transmitted down the central canal can cause syringomyelia.[11,49] Syringomyelia is commonly associated with ACM-1 in as many as 70–80% of the cases.[4,14,22] ACM-1 with syringomyelia can develop into progressive neurological deterioration. Nonetheless, the size of the syrinx can diminish following appropriate ACM surgical correction.[26,28,45] ACM-1 is also commonly associated with scoliosis, kyphosis, hydrocephalus, platybasia, occipitalization, or basilar invagination.[21,37]

Increased frequency in magnetic resonance imaging (MRI) use has revealed a higher incidence of ACM-1 than initially thought, accounting for 0.6–0.7% of the cases.[31]

Symptomatology

ACM-1 presents with different degrees of neurologic manifestations that can be evident during the second or third decades of life. The most commonly reported symptoms are suboccipital headache (81%), occular manifestations (78%), otoneurologic disturbances (74%), pain in the extremities, cranial nerves disturbances (hoarseness, cough, dysphagia), weakness and muscle atrophy, and sensory disturbances (numbness, pain, and temperature deficits).[10,32] Often headaches originating from ACM-1 can be confused with other types of headaches; however, the typical description of the headaches in ACM-1 are described by patients as pressure-like occipital headache, radiating to the vertex and behind the eyes, and sometimes down the neck and into the shoulders.[32] Meadows *et al.* retrospectively analyzed MR images of the head and cervical spine in 22591 patients, and 175 patients were found to have ACM-1 with tonsillar herniation more than 5 cm below the foramen magnum.[31] Out of the 175 patients with ACM-1, 25 were clinically asymptomatic and one of

them had reported a headache that was ruled out by the neurologist as an ACM-1 related symptom. The average degree of herniation in the asymptomatic patient group was 11.4 ± 4.86 mm.[31]

Clinical diagnosis

MRI or computed tomography (CT) scan are usually sufficient to determine the position of cerebellar tonsils, abnormalities in the posterior fossa, and the presence of syringomyelia.[5,7,12] MRI can determine the severity of ACM-1 and the effect it has on the CSF circulation.[13] Based on the radiologic findings, ACM-1 is defined as herniated cerebellar tonsills >5 mm in adults and >6 mm in children.[13] When the tonsillar herniation is determined to be between 3–5 mm, it can be defined as a benign cerebellar ectopia which requires close monitoring and correlation with the patient's symptomatology.[7] Interestingly, approximately 30% of a patient population with a 5–10 mm cerebellar tonsillar displacement will have no symptoms.[2]

Another important component of the ACM-1 is the disturbance in CSF dynamics which can be assessed through CSF fluid flow MRI.[29] The velocity of the CSF flow through the foramen magnum depends on the dimensions and the crowding of the foramen magnum.[16] Patients with ACM-1 present with increased velocity of CSF flow (peak systolic velocity of 3.1 cm/s and peak diastolic velocity of 4.0 cm/s); nonuniform flow secondary to mechanical obstruction during the cardiac cycle: Anterior and posterior flow disparity; and simultaneous bidirectional flow.[13,17]

On imaging studies, syringomyelia is commonly detected at the C4-C6 spinal cord levels, however, it can involve the entire length of the spinal cord.[13,40]

Obstetric concerns in ACM-I

ACM-1 is of a significant clinical concern in pregnant patients, first because of physiological increase in CSF pressure associated with pregnancy, and second because delivery represents a vulnerable period that puts parturients at a great risk of complications due to an acute increase in intracranial pressure (ICP). The findings of the Meadows *et al.* study alerts us to the fact that an advanced form of ACM-1 with minimal symptoms or advanced asymptomatic ACM-1 can lead to catastrophic consequences during labor and delivery.[18,31,42]

Some authors have suggested that "pushing" during labor increases CSF pressure by approximately 20–51 mmHg; however, Marx *et al.* suggested that the increase in CSF pressure occcurs due to the pain experienced by the patient during uterine contractions.[25,27]

Throughout the course of the first stage of labor, when the cervix effaces and dilates (early labor), and uterine contractions are becoming longer, stronger, and closer together (active labor), it was found that the basal

CSF pressure was only 13 mmHg ± 2.5 mmHg after eliminating the pain factor.[19]

During the second stage of the labor (full cervical dilatation followed by delivery of the baby) and third stage (delivery of placenta), where most of the "pushing" takes place, the CSF pressure increases, on average, by 8 mmHg or by 20–25% at the time of delivery.[19]

Although some authors state that, it is not known whether uterine contractions and "pushing" during labor worsen the ACM-1 condition or not, the obstetric and anesthesiologic management of such patients remains a dilemma.[18,42]

Despite the lack of evidence-based guidelines and no uniform recommendations regarding management of these patients throughout pregnancy and delivery, a multidisciplinary approach is implemented based on best clinical judgement and in-hospital protocols. We present a case of ACM-1 where the importance of a multidisciplinary team approach, including anesthesiologists, neurosurgeons, and obstetricians can actively intervene in the diagnostic process and mode of delivery to prevent possible severe morbidity and mortality.

CASE DESCRIPTION

A 24-year-old, ASA II, G1P0 full-term parturient presented to the Labor and Delivery for vaginal delivery. Her past medical history was significant for gestational diabetes and ACM-1.

A diagnosis of ACM-1 was made 12 years ago when a brain MRI was performed for right-sided numbness developing following a motor vehicle collision. The patient had been entirely asymptomatic since then, and had been seen by an outside neurologist frequently for the past 10 years. She was assured by her neurologist 2 months prior that vaginal delivery and labor epidural placement would be safe, however she had not been seen by an anesthesiologist in the anesthesia preop clinic before.

In the 39th week of pregnancy, she presented to our hospital for rupture of membranes and uterine contractions; anesthesia was consulted for an epidural placement. During the anesthesia evaluation, it was noted that she had an exaggerated patellar reflex and a questionable left-sided Babinski; subsequently, MRI images were requested from an outside hospital. A thorough review of the brain MRI demonstrated an advanced form of ACM-1 with 1.7 cm transtonsillar herniation and a large syrinx extending down from C1 to C5. Concerned with the progression of the transtonsillar herniation during labor, an urgent neurosurgery consult was requested. Following a discussion with the patient,

family, primary OB team, and the neurosurgeon, a plan for cesarean section was made per neurosurgical recommendations, including avoidance of neuraxial anesthesia and use of careful laryngeal manipulation and endotracheal intubation.

A bolus dose of terbutaline was given to stop active contractions and the patient was brought to the operation room. The patient was placed in a left lateral uterine displacement position, and standard ASA monitors were applied. Concern regarding further progression of the herniation due to increased ICP during head and neck movement and laryngoscopy, led to an awake fiberoptic intubation was performed under generous topical anesthesia consisting of 6 ml of 1% lidocaine spray. A multimodal general anesthesia was used with the mixture of 1 MAC of desflurane, oxygen, and air and controlled ventilation. Mild hyperventilation was provided to ensure an end-tidal CO_2 of 30 mmHg to prevent a possible increase in ICP. A healthy infant male was delivered with Apgar scores at 1 and 5 minutes of 9/9. After umbilical cord clamping, an infusion of mannitol 0.5 mg/kg and dexamethasone 10 mg was given.
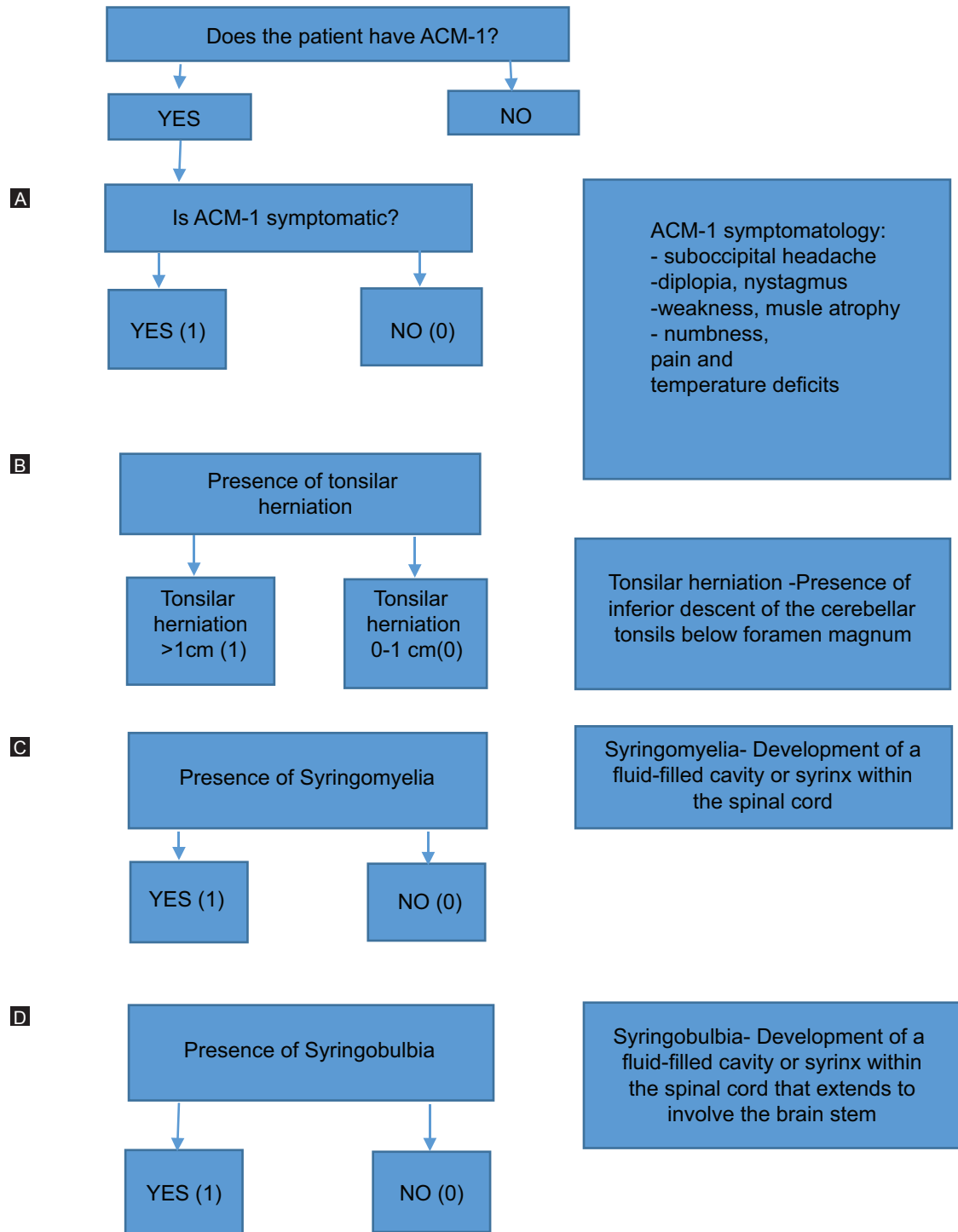
The remainder of the case was without incident. Post-extubation neurological assessment was grossly normal. The patient had no complaints of headache or pain in the post-anesthesia care unit and she was advised to follow-up with the neurosurgeon later.

DISCUSSION

The case described above reflects an example of unfamiliarity of the consultant neurologist with regards to the management of parturients with ACM-1, leading to an underestimation of the patient's ACM-1 severity, and assuring the patient that vaginal delivery with an epidural anesthetic is safe. Therefore, the first author, based on his dual training, provided a thorough understanding of the physiology of ACM-1 in terms of the neurosurgical and anesthesiology concerns. Therefore, he was able to establish a correct diagnosis by personally interpreting the patient's MRI.

Based on this case report and other similar cases found in the literature where the obstetricians found themselves alone with an ACM-1 patient, the first author developed a questionnaire to utilize when faced with such cases [Figure 1].

In order to prevent the occurrence of a preventable catastrophe, the obstetric specialists need to raise awareness of the ACM-1 condition in pregnant patients to their primary care provider and specialists and vice-versa. In addition, a thorough examination of the most recent MRI with the patient, anesthesia provider, and a neurospecialist should be performed prior to deciding on the best mode of delivery. The first author has developed

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                    ┌─────────────────────────────────┐
                    │   Does the patient have ACM-1?   │
                    └─────────────────────────────────┘
                         │                      │
                    ┌─────────┐           ┌─────────┐
                    │   YES   │           │   NO    │
                    └─────────┘           └─────────┘
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A

```
┌─────────────────────────────┐
│     Is ACM-1 symptomatic?    │
└─────────────────────────────┘
        │              │
  ┌──────────┐   ┌──────────┐
  │ YES (1)  │   │  NO (0)  │
  └──────────┘   └──────────┘
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ACM-1 symptomatology:
- suboccipital headache
-diplopia, nystagmus
-weakness, musle atrophy
- numbness,
pain and
temperature deficits

B

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┌─────────────────────────────┐
│    Presence of tonsilar      │
│         herniation           │
└─────────────────────────────┘
        │              │
  ┌──────────┐   ┌──────────┐
  │ Tonsilar │   │ Tonsilar │
  │herniation│   │herniation│
  │ >1cm (1) │   │0-1 cm(0) │
  └──────────┘   └──────────┘
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Tonsilar herniation -Presence of inferior descent of the cerebellar tonsils below foramen magnum

C

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┌─────────────────────────────┐
│   Presence of Syringomyelia  │
└─────────────────────────────┘
        │              │
  ┌──────────┐   ┌──────────┐
  │ YES (1)  │   │  NO (0)  │
  └──────────┘   └──────────┘
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Syringomyelia- Development of a fluid-filled cavity or syrinx within the spinal cord

D

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┌─────────────────────────────┐
│   Presence of Syringobulbia  │
└─────────────────────────────┘
        │              │
  ┌──────────┐   ┌──────────┐
  │ YES (1)  │   │  NO (0)  │
  └──────────┘   └──────────┘
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Syringobulbia- Development of a fluid-filled cavity or syrinx within the spinal cord that extends to involve the brain stem

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┌──────────────────────────────────────────────────────────────────┐
│   1 point - multidisciplinary meeting on the site                  │
│                                                                    │
│  2 points- neurosurgery consult and multidisciplinary meeting on   │
│  the site                                                          │
│ >3 points- neurosurgery consult and preparation for C-section      │
└──────────────────────────────────────────────────────────────────┘
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Figure 1: Ghaly Obstetric Guide to Arnold-Chiari malformation type 1 (GOGAC-1)

a set of guidelines [Table 1] that describes the most crucial steps required to be followed perioperatively with cases involving pregnancy and ACM-1.

This case demonstrates how the anesthesia team was instrumental in changing the course of labor and delivery by using a comprehensive physical examination and a thorough review of the brain MR imaging, thus helping prevent possible maternal-newborn morbidity and mortality.

It is paramount to review the MRI report, as well as to independently interpret the images, searching for the position of the tonsils and the presence of a syrinx while correlating with the patient's symptomatology. Neurosurgical consult has to be available and performed before proceeding with the delivery. It is critical that the anesthesiology specialist has proper training regarding the specificities of ACM-1 and can intervene in the diagnosis and peripartum management of this classification of patients.

Pathophysiology of syringomyelia and syringobulbia

Syringomyelia presents in two forms, namely, noncommunicating and communicating.[33] The communicating form is described as a continuity between the syrinx and the CSF in the central canal of the spinal cord. It is the most common form, which is also associated with ACM-1.[33] Occurrence of a craniospinal pressure gradient is suggested to be associated with the propagation of pressure waves into the central canal and subsequent dissection into adjacent neural tissue.[47] The

syrinx is commonly located in the lower cervical or upper thoracic region and gradually expands into the anterior and lateral cords or upwards into the medulla with the development of syringobulbia. The cystic cavities of syringomyelia can damage the white and gray matter of the spinal cord, with further involvement of fibers subserving sensation of pain and temperature.[47] In addition, the syrinx can destroy first and second order motor neurons, leading to flaccid weakness in the upper extremities and spastic paresis in the lower extremities. Other symptoms are hyperhidrosis, loss of reflex bladder emptying, decreased respiratory drive, impaired vagal cardiovascular reflexes, etc.[24,36]

The main goal of anesthesia management in parturients with ACM-1 and presence of syringomyelia is to avoid increase in craniospinal pressure. An increase in this gradient or ICP can lead to the herniation of the cerebellum/brainstem or further extension of the syrinx. Avoiding increases in intrathoracic pressure, such as with sneezing, coughing, vomiting, and Valsalva maneuver, is vital, and thus Cesarean section under general anesthesia is recommended for delivery.[9,33]

Syringobulbia can occur in a previously established syringomyelia due to upward pulsatile fluid movements.[34] There are hypotheses that syringobulbia in ACM-1 can occur in the presence of craniocervical bony malformations.[46] Syringobulbia can be represented by trigeminal, vagus, oculomotor, abducens, facial, glossopharyngeal, and hypoglossal nerve dysfunctions. Some other symptoms may include neck pain, syncope, vertigo, weakness, and sensory dysfunction.[30,41,47,48]

Management of parturients with ACM-1 during delivery

General anesthesia in ACM-1 parturients
General anesthesia avoids the risk of dural puncture in patients with increased ICP. It also can help manage the sudden increases in intracranial pressure intraoperatively by hyperventilating the patient, keeping the airway secure, and controlling blood pressure.[6,15]

Chantigian *et al.* conducted a retrospective study on parturients from their institution diagnosed with ACM-1 and the mode of delivery.[6] Out of 14 patients that were known at the time of delivery to have ACM-1, 10 underwent vaginal delivery and 4 cesarean deliveries. Out of the 10 patients that delivered vaginally, 8 received perineal analgesia with inhalation anesthesia. General anesthesia was also the anesthetic management for one parturient who delivered via cesarean section. None of these patients developed new symptoms or worsening of their preoperative symptoms of ACM-1.[6]

Despite such promising results, laryngeal manipulation and endotracheal intubation can lead to abrupt increases in ICP with serious consequences; for this reason, some authors prefer awake intubation with

Table 1: Ghaly general anesthesia recommendations for cesarean section in parturients with ACM-1

Notify Neurosurgery service and open communication

Anesthesia time-out, add ACM-1 concerns to the surgery standards time out

Avoiding blood pressure fluctuation and ICP elevation with measures to reduce the ICP throughout the procedure

Prophylactic antacid protocol

ASA monitors and preoxygenation

Simultaneous surgical field preparation

Adequate topicalization and awake fiberoptic intubation

Consider hyperventilation and osmodiuresis

Post-intubation neurological assessment

Propofol IV 1 mg/kg for induction followed by balanced anesthesia with volatile anesthetic, Sevoflurane 0.5 MAC and additional IV infusions of Propofol and Remifentanil

Postdelivery osmodiuresis (Lasix 0.5 mg/kg, Manitol) and hyperventilation (CO_2 30-35 mmHg)

Immediate delivery of the baby

Ensure smooth emergence: avoid increase in ICP

Confirm full NMB reversal

Prior extubation neurological assessment

Standby neurosugeon on call

local airway anesthesia to prevent serious complications from occuring,[15] whereas others prefer laryngoscopy intubation using a modified rapid sequence technique, and opioids.[39] Another concern with general anesthesia for parturients with ACM-1 is the presence of difficult airway that can be underestimated on routine preoperative examination. Therefore, it is recommended to anticipate complications of this nature and take the required steps in preventing hypoxia and damage to the spinal cord.[35]

Parturients that present with ACM-1 and syringomyelia have an increased sensitivity to neuromuscular blocking agents; some anesthesiologists prefer to avoid them altogether, whereas others closely monitor neuromuscular function for prolonged effects and then provide complete reversal.[15]

Spinal anesthesia in ACM-1 parturients
Two cases reported in the literature demonstrated mild-to-more severe complications following spinal anesthesia performed in two parturients with ACM-1.[6,20] One parturient developed progressive postural headache associated with visual changes and nausea that resolved with an autologous epidural blood patch, however, the symptoms recurred and intensified days later. Resolution occured only after 6 weeks of prednisone therapy. Another parturient, who was managed with continuous spinal anesthesia, developed postdural puncture headache that was subsequently resolved successfully with epidural blood patch. These two cases warn of the serious consequences that could occur following subarachnoid anesthesia in parturients with known or more concerning unknown ACM-1.[3,20]

Epidural anesthesia for parturients with ACM-1
Epidural anesthesia is preferred over general anesthesia in some instances in parturients with ACM-1 during delivery. It is worth mentioning that, in the nonpregnant population with an elevated baseline ICP, a transient increase of ICP occurs after epidural injection of 10 mL bolus of local anesthetic given over 20–30 s that lasts for 4.5 min (average increase of 21 mmHg) (from 18.8 to 39.5 mmHg). In comparison, in the nonpregnant population with pre-injection normal ICP, there was an average 6 mmHg (9.3–15.6 mmHg) increase in ICP that lasted for 2.3 min. Reduction in the volume of bolus to 5 mL in patients with an elevated ICP significantly reduced the increase in ICP to 5 mmHg for a period of 2.8 min.[18]

Semple *et al.* reported an uncomplicated cesarean delivery conducted under epidural anesthesia with incremental local anesthetic boluses followed by continuous infusion in a patient with ACM-1.[42] In addition, Penney *et al.* reported an uneventful vacuum-assisted vaginal delivery in a 30-year-old parturient, who was managed with epidural anesthesia with small divided doses of bupivacaine 0.2% followed by a continuous infusion of 10 mL/h bupivacaine 0.1% and fentanyl 2 mcg/mL.[39]

One parturient developed severe complications following unintentional dural puncture during epidural anesthetic placement and demonstrated symptoms of episodic headaches and progression to gait instability 1 year later.[3]

Although successful deliveries were acomplished with this method of anesthesia, the potential risks of inadvertant dural puncture during needle or catheter insertion can lead to serious complications. In order to prevent them it is recommended to use small bolus doses of local anesthetic medications, which would have less effect on increasing ICP in susceptible patients.

Other ACM-1 particularities
In patients with known ACM-1, comprehensive neurologic evaluation should be conducted before deciding about the mode of delivery, and again after delivery to look for any signs of worsened neurological symptoms or evolution of new symptoms. Neurological assessment needs to include identification of autonomic neuropathy, especially cardiac autonomic neuropathy as it can present with tachiarrhythmias and wide fluctuations in arterial blood pressure intraoperatively. Furthermore, sudden cardiac death and respiratory arrest were reported in patients with syringomyelia and concomitant diabetic neuropathy following general and regional anesthesia.[23,38] Another aspect to consider in ACM-1 is the association with kyphosis before proceeding with general anesthesia, which requires performance of pulmonary function tests, which can demonstrate flattening of the inspiratory curve on the flow-volume loop.[41]

CONCLUSION

Unfamiliarity of health care providers with regard to ACM-I parturients can be countered with increasing awareness to this condition throughout the medical specialties involved in their care. The Ghaly Obstetric Guide to Arnold-Chiari malformation type 1 (GOGAC-1) can assist in making the right decision when faced with such cases. The GOGAC-1 guideline is an excellent tool in aiding with the process of taking the next best step in the management and diagnosis for providers, especially for those who come in contact with such patients for their first time.

Depending on the degree of ACM-1 severity, the Ghaly Obstetric Guide to ACM-1 dictates that a multidisciplinary approach determines the mode of delivery. Correct interpretation of the imaging tests with thorough neurological, cardiac, and respiratory examination should be conducted before making a decision on anesthetic management. The table developed by the senior author regarding the recommendations to be followed for ACM-1 parturients perioperatively

helps prevent and manage potential complications. Furthermore, postoperative pain should be addressed in such patients; thus, a preemptive discussion on the best options available for pain management has to take place between specialists involved in the care of the patient.

These steps along with proper training of the anesthesia providers regarding the specificities of ACM-1 parturients aids in better management and understanding of this complex condition.

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Conflicts of interest
There are no conflicts of interest.

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