BIOLOGY AND MOLECULAR MEDICINE

Case Report

Anesthetic Management of Symptomatic Chiari I Malformation in a Parturient: A Case Report

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Abstract

Anesthetic management of parturients with symptomatic Chiari I malformation requires individualized assessment, as both neuraxial and general anesthesia carry theoretical risk. With regard to neuraxial anesthesia, unintentional dural puncture may produce a cerebrospinal fluid pressure gradient, which can result in further neurologic deterioration. Additionally, manipulations involved in administration of general anesthesia, including direct laryngoscopy and endotracheal intubation, without optimal medication, can result in a hypertensive response and increased intracranial pressure. We report the case of a 25-year-old parturient with a Chiari I malformation, with associated progressive neurological symptoms, and a 14-mm cerebellar tonsillar descent, who underwent cesarean delivery under general anesthesia following multidisciplinary consultation. Targeted pharmacological and hemodynamic strategies were implemented to blunt sympathetic responses during airway management. Deep extubation was complicated by laryngospasm, requiring re-sedation and re-administration of a paralytic with positive-pressure bag-mask ventilation, after which the patient was neurologically at preoperative baseline. This case illustrates the competing neuraxial and general anesthetic risks involved in managing patients with symptomatic Chiari I malformation during pregnancy.

Keywords: Chiari I malformation, Neuraxial Anesthesia, Intracranial Pressure

Abbreviations: Cerebrospinal fluid (CSF), Intracranial pressure (ICP), Magnetic resonance imaging (MRI)

1. Background

Chiari I malformation is a hindbrain maldevelopment characterized by downward displacement of cerebellar tonsils into the foramen magnum of at least three to five millimeters. The presenting symptoms and signs may range from headache and blurry vision to unsteady gait and proprioceptive deficits [1, 2].

There are numerous reports in the literature of successful neuraxial anesthesia delivery in patients with Chiari malformation, including patients with and without known diagnoses [3]. A retrospective chart review by Chantigian et al. assessed anesthetic outcomes in 12 patients with Chiari I malformation who delivered 30 babies. Epidural anesthesia was used for six deliveries, spinal anesthesia for two, a continuous spinal catheter for one, and general anesthesia for three cesarean deliveries. The patient who received continuous spinal anesthesia developed a postdural puncture headache responsive to an epidural blood patch. No exacerbation of pre-existing neurologic symptoms was reported [4].

Currently, anesthetic management of parturients with known Chiari I malformation remains unclear and controversial, with the potential for poor outcomes. Leffert and Schwamm offer a review and assessment of numerous risks and pitfalls of various strategies in anesthetic management [3]. With regard to neuraxial anesthesia, an unintentional dural puncture may produce a cerebrospinal fluid (CSF) pressure gradient between the brain and spinal cord, which can result in further displacement of brain tissue and worsened neurologic function [3, 5]. Additionally, manipulations involved in administration of general anesthesia, including direct laryngoscopy, endotracheal intubation and extubation, without optimal medication, can result in a hypertensive response and increased intracranial pressure (ICP). An increased ICP can exacerbate the CSF pressure gradient, resulting in the downward displacement of brain tissue. A smooth rapid sequence

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induction with pharmacologic blunting of sympathetic responses to endotracheal intubation and extubation should be implemented. Moreover, bucking and coughing upon extubation should be minimized, as this can also increase ICP. The practice of extubating under deep anesthesia decreases the aforementioned risks and is a suitable option to mitigate harmful effects of increased ICP with the understanding that protective airway reflexes have not yet returned, and the risk of aspiration of gastric contents is increased [6, 7].

We report the case of a parturient with a symptomatic Chiari I malformation with a 14-mm cerebellar tonsillar descent, with cervical and thoracic syrinxes who underwent cesarean delivery under general anesthesia. The case highlights individualized choice of anesthetic technique, efforts to limit airway-related increases in intracranial pressure, and laryngospasm during attempted deep extubation.

2. Case Presentation

A 25-year-old parturient, gravida three para two, with a history of asthma, chronic hypertension, deep vein thrombosis, and Chiari I malformation, presented to labor and delivery at 30 weeks and 6 days gestation due to worsening headaches, blurred vision, and unsteady gait.

The patient's neurologic history extended back six years, with headaches during both previous pregnancies. In 2017, her first pregnancy, she was admitted to the hospital with a diagnosis of pre-eclampsia with severe features due to headaches. The presence of Chiari I malformation was unknown at the time, and she received a labor epidural. Labor was induced, resulting in a spontaneous vaginal delivery at 32 weeks and 0 days. In 2021, her second pregnancy, she was admitted to the hospital due to an asthma exacerbation, and was then diagnosed with pre-eclampsia with severe features due to headaches. A brain MRI, her first known imaging, revealed a Chiari I malformation with cerebellar tonsils 8.5 mm below the foramen magnum. She underwent cesarean section delivery with successful spinal anesthesia at 24 weeks and 1 day, complicated by neonatal demise on postoperative day two due to intraventricular hemorrhage.

The patient's headaches progressed from occurring during pregnancy only, to intermittent, occurring two to three times weekly following her second pregnancy. Her headaches were described as holocephalic, more severe over the temples, pulsatile, moderate to severe intensity, and associated with photophobia, without nystagmus, diplopia, or ataxia. A second MRI, approximately two years following the first MRI, revealed peg-shaped cerebellar tonsils descending 14 mm below the foramen magnum, causing crowding of the cervicomedullary junction (Figure 1). Two areas of syrinx were found in the spinal cord at levels C5–C6 and T1–T2, measuring 2.2 mm and 2.4 mm respectively in the craniocaudal direction. The cervical and thoracic syrinxes demonstrated spinal cord involvement in the setting of her Chiari I malformation.

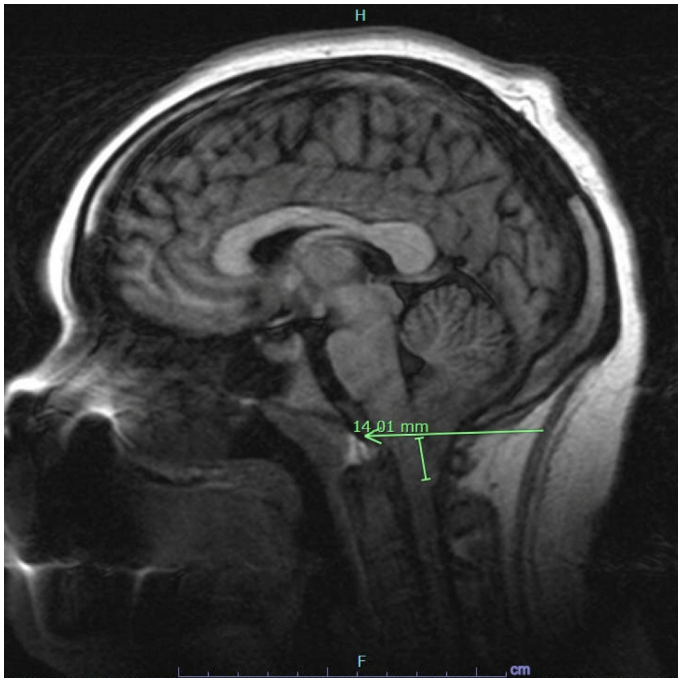

Figure 1. Peg-shaped cerebellar tonsils descending 14 mm below the foramen magnum, causing crowding of the cervicomedullary junction.

During the third pregnancy in 2023, the patient was admitted to the hospital at 30 weeks and 6 days due to worsening headaches. She described constant headaches associated with diplopia, blurry vision, and unsteady gait. Proprioceptive defects were noted in the upper and lower extremities bilaterally, with associated Romberg sign, and nystagmus. Given the patient's progressive neurologic symptoms, more severe in this current pregnancy than prior, a 14-mm cerebellar tonsillar descent, and syringomyelia, following multidisciplinary discussion including Obstetrics, Neurology, Neurosurgery, and Anesthesiology, it was advised that the risk of neuraxial anesthesia would outweigh its benefit, and continuation of the pregnancy with progressive neurologic deterioration with eventual active labor put the patient at risk for worsening neurologic status and potential permanent disability. Therefore, general anesthesia for cesarean section was selected.

The patient underwent cesarean section at 31 weeks and 5 days under general anesthesia. Midazolam (2 mg) was given preoperatively, and radial arterial catheterization was performed. A nicardipine intravenous infusion at a rate of 5 mg/h was started preinduction. For intravenous induction of general anesthesia, 200 mg of propofol, 250 µg of fentanyl (2 µg/kg of total body weight of 127 kg), and 100 mg of rocuronium were administered. At the time of endotracheal intubation, two 10-mg boluses of labetalol were administered. Direct laryngoscopy and endotracheal intubation were well tolerated; the airway was secured uneventfully, and invasive blood pressure measurement peaked at 130/53 mmHg at that time. Sevoflurane was used for maintenance of anesthesia. The neonate was delivered eight minutes following intubation. During attempted extubation under deep general anesthesia, the patient experienced laryngospasm, and the decision

was made to deepen the anesthetic and to re-administer a short-acting paralytic. 50 mg of propofol and 40 mg of succinylcholine were administered to provide rapid-onset sedation and paralysis. The patient remained extubated and positive-pressure mask ventilation was administered for 21 minutes until re-emergence from anesthesia. At that time, the patient was hemodynamically stable, without coughing, and neurologically at preoperative baseline on examination.

Three weeks following cesarean section, the patient underwent elective suboccipital craniectomy with C1 laminectomy for Chiari decompression. The principal clinical findings and perioperative course are summarized in Table 1.

Table 1. Clinical summary: Symptomatic Chiari I malformation during cesarean delivery

Category	Clinical summary
Patient	25-year-old, gravida three para two, with a history of chronic hypertension, asthma, deep vein thrombosis, and Chiari I malformation; underwent cesarean delivery at 31 weeks and 5 days.
Neurologic findings	Progressive headaches, diplopia, blurred vision, unsteady gait, proprioceptive deficits, Romberg sign, and nystagmus.
Imaging	Tonsillar descent measured 8.5 mm on initial MRI and 14 mm on later MRI, with cervicomedullary crowding. Syrinxes were present at C5–C6 and T1–T2.
Anesthesia	General anesthesia selected after multidisciplinary discussion in the setting of progressive symptoms, 14-mm tonsillar descent, and syringomyelia.
Perioperative care	Radial arterial catheter and nicardipine infusion before induction; propofol, fentanyl, and rocuronium for induction; labetalol at intubation and sevoflurane for maintenance. Peak recorded blood pressure at intubation: 130/53 mmHg.
Emergence	Deep extubation was complicated by laryngospasm. Propofol and succinylcholine were given, followed by positive-pressure mask ventilation and re-emergence.
Outcome and unavailable data	On emergence, she was hemodynamically stable, without coughing, and neurologically at preoperative baseline. Underwent Chiari decompression three weeks later. Perioperative ICP measurements, detailed neonatal outcomes, and long-term neurologic follow-up were unavailable to the authors.

3. Discussion

Safe anesthetic management of parturients with known Chiari I malformation poses unique challenges. The chief concern with neuraxial anesthesia, spinal or accidental dural puncture during attempted epidural placement, is the creation of a CSF pressure gradient between the brain and spinal cord, which can result in further displacement of brain tissue and worsened neurologic function [3, 5]. However, general anesthesia also carries risk, due to sympathetic surges during direct laryngoscopy, endotracheal intubation and extubation that can transiently increase intracranial pressure and result in further neurologic deterioration [6].

In our reported case, deep extubation was utilized to minimize coughing or bucking against the endotracheal tube during emergence, which would transiently increase ICP. Although remifentanil can blunt coughing reflexes during awake extubation, it can result in respiratory drive depression, causing hypoventilation and subsequent hypercapnia, which would raise ICP [8]. Concerns of increased ICP outweighed concern for aspiration in the decision to extubate under deep anesthesia.

Available retrospective studies and reviews suggest that neuraxial anesthesia may be used with low reported rates of neurological complication in carefully selected patients with Chiari I malformation, including some patients with existing syringomyelia. Garvey et al. reported no maternal or neonatal complication associated with neuraxial anesthesia in their case series and review of syringomyelia in pregnancy [9]. Gruffi et al. found that anesthetic complications occurred infrequently, most commonly postdural puncture headache [5]. A 2025 retrospective analysis by Pulido et al. evaluated 76 deliveries in 36 patients, including 60 neuraxial anesthetics, with Chiari I malformation, with and without associated syrinx and found no significant worsening of neurologic symptoms or MRI findings [10].

The 2026 Obstetric Anaesthetists' Association multidisciplinary consensus statement concluded that most patients with existing Chiari I malformations with stable or no neurologic symptoms can be managed with routine obstetric anesthesia protocols after careful assessment. However, the group only commented on patients without significant or unusual Chiari-related symptoms [11]. Their consensus statement strengthens our rationale for reporting this rare and unusual clinical case.

Parturients with Chiari I malformation present a challenge regarding anesthetic management as both neuraxial and general anesthesia carry inherent risks in this patient population. To date, there are no published randomized controlled trials comparing parturients with intracranial pathology undergoing neuraxial anesthesia to those receiving a general anesthetic. A multidisciplinary approach involving Obstetrics, Neurology, Neurosurgery, and Anesthesiology is recommended to facilitate individualized planning and to mitigate potential neurologic and obstetric complications.

4. Conclusion

We report the case of a 25-year-old parturient with progressive, symptomatic Chiari I malformation who successfully underwent cesarean section under general anesthesia. Following multidisciplinary assessment, general anesthesia was selected with targeted pharmacological and hemodynamic strategies to blunt sympathetic responses during airway management. Although deep extubation was complicated by laryngospasm, the patient remained neurologically at baseline at the last documented postoperative assessment. This case highlights the importance of neurologic assessment, careful anesthetic selection, and multidisciplinary individualized planning.

Prior Presentation: This case report was presented at the Society for Obstetric Anesthesia and Perinatology (SOAP) 2024 meeting.

Limitations: Detailed neonatal outcome data and longer-term neurological follow-up after decompression were not available to the authors. Consequently, the report cannot assess neonatal anesthetic outcomes or maternal recovery beyond the final documented follow-up.

Informed Consent Statement: Written informed consent was obtained from the patient for publication of this case report and the accompanying clinical information and image.

Institutional Review Board Statement: In accordance with the policy of Trinity Health of New England, this retrospective single-patient case report did not constitute human-subjects research and did not require Institutional Review Board review or approval.

Data Availability Statement: All relevant clinical information is included in the article. Additional patient-level data are not publicly available because of patient privacy restrictions.

Author Contributions: K.R. wrote and prepared the case report for submission. C.C. was the resident physician for the operative case and edited the case report. J.Z. provided expertise on the existing literature on the subject matter. R.Z. was the attending anesthesiologist for the case, who provided preoperative and intraoperative anesthetic care. All authors have agreed to the publication of this case report.

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