Management of Emergent Cesarean Section for Twin B in Parturient with Arnold Chiari I Malformation

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Introduction: Arnold Chiari malformations (ACM) are congenital disorders classified by the degree to which cerebellar tonsils and brainstem structures are herniated through the foramen magnum. ACM type I are the most common in adults and associated with syringomyelia1,2. Questions still exist regarding the best way to manage the parturient with ACM 1. Although there have been case studies detailing the management of laboring patients with Chiari malformations, none has discussed the management of twin deliveries. The largest report by Chantigan3 described 12 parturients over a 50-year period with ACM I, some of which were diagnosed postpartum. All underwent uneventful general anesthesia as well as neuraxial anesthesia without complications. Landau4 discusses several case reports and series that have been published regarding the safety of epidural and spinal anesthesia in ACM I. However, there are concerns when administering neuraxial anesthesia which include a risk of further herniation and exacerbation of symptoms. No articles to date have been published detailing management of emergent cesarean section with a functioning epidural or an increased risk for high epidural as a potential complication related to neuraxial anesthesia.

Case Description: 37-year-old female G1P0 with ACM 1, GDMA1, chronic HTN with superimposed preeclampsia without severe features presented with dichorionic, diamniotic twins gestation at 34 weeks requesting labor analgesia. She had no prior surgical interventions for ACM 1 and was neurologically intact without symptoms or signs of increased ICP. She underwent uneventful placement of a combined spinal and patient controlled epidural. She received fentanyl 25 mcg intrathecally and 3 cc lidocaine 1.5% with epinephrine epidural test dose via catheter. Incremental dosing at 5 and 12 minutes later with 4 cc of 0.25% bupivacaine resulted in a T4 sensory level with minimal hemodynamic changes. A fentanyl 2 mcg/mL and bupivacaine 0.125% infusion was started at 8 mL/hr. Thirty-five minutes after epidural placement, fetus A experienced bradycardia. The patient was completely dilated and transported to the operating room for an attempt at vaginal delivery. Baby A was delivered vaginally with the assistance of forceps. Baby B, who was in a breach position with a high station, experienced significant bradycardia. The decision was made to proceed with emergent cesarean section. 20 cc of 2-chloroprocaine 3% was bolused at 55 minutes after epidural placement and C section was performed with adequate analgesia. Baby B was uneventfully delivered. Thirteen minutes after chloroprocaine was bolused, the patient complained of difficulty breathing. She had a T1 sensory level with profound hypotension (MAP 40s-50s) and bradycardia (HR 50s) which required multiple boluses of phenylephrine and ephedrine, eventually necessitating a phenylephrine infusion. She was placed in the reverse Trendelenburg position and a frequent check of sensory level. At the conclusion of the case, the patient was able to move herself
to the hospital bed. 1.5 hours after the chloroprocaine bolus, sensation and strength were intact. The remaining of the patient’s recovery was uneventful. She suffered no neurologic sequelae.

Discussion: Although neuraxial anesthesia is considered safe in asymptomatic patients with ACM 1, management may still pose a challenge. Sicuranza et al recommend avoidance of spinal anesthetics in symptomatic patients due to potential for increased ICP after lumbar puncture and exacerbation of the malformation especially in those with signs of increased ICP. Furthermore, they warn against the potential for increased ICP with epidural anesthesia especially with large boluses in rapid succession. Epidural anesthesia does increase the risk of tentorial herniation, potentially lead to a decrease in brain perfusion with hypotension, and with an accidental wet tap may lead to exacerbation of brain fluid shifts. Benefits of epidural anesthesia may include a decrease in ICP from improved pain control and improved patient experience. Choi et al endorse careful weighing of the previously discussed risks versus benefits of epidural anesthesia on a case by case basis. In a case report, Nel describes successful and careful incremental dosing of bupivacaine in a patient with ACM-I and syringomelia for a cesarean section. However, there is no available data on how the anesthesiologist should proceed with patients, who already have a functioning epidural placed for labor analgesia, in the case of an emergency cesarean section.

Most authors suggest that general anesthesia is preferred in patients with signs of increased ICP undergoing cesarean section. Due to the ability to control increases in ICP with hyperventilation, maintaining a secure airway, and tighter blood pressure control, general anesthesia in patients with symptomatic ACM-1 and signs of increased ICP should be strongly considered whether emergent or otherwise. General anesthesia is not without risk in obstetric patients even without ACM-I due to their proclivity for difficult airways, increased risk of aspiration, and decreased FRC leading to rapid desaturation. These parturients are challenging due to their need for RSI for a presumed full stomach and avoidance of neck hyperextension, which could lead to brainstem compression and stretching of cranial nerves. However, it is unclear whether a patient undergoing an emergent caesarian section with an asymptomatic ACM should receive general anesthesia in hopes of avoiding the risk of increasing her ICP, possible exacerbation of ACM, and even tentorial herniation. Without signs of increased ICP, epidural anesthesia with incremental boluses is preferred if there is sufficient time according to Landau’s review.

Even the preferred mode of delivery is a topic of controversy. Vaginal delivery comes with the risk of increased ICP from both pain and uterine contractions. Although, it is uncertain if this increase in ICP leads to a clinically significant outcome based on studies performed in the 1960s, cesarean sections themselves are associated with significant surgical risks as well as anesthetic concerns previously mentioned.

One of the anesthesiologist’s concerns regarding Chiari malformations is inducing further herniation by inadvertent wet tap or dural puncture with a large bore needle. Epidural block spread is more pronounced with increased volume of local anesthetic, advanced age, decreased height, pregnancy, and decreased epidural space, e.g. spinal stenosis. High epidurals due to increased epidural pressure or decreased epidural volume are not typically associated with this disorder. There are no other case reports describing occurrence of high epidurals in patients with ACM and further studies remain necessary determine relation between high epidurals and ACM. We hypothesize that the high epidural is related to the amount of local anesthetic administered over
a relatively short time frame from epidural placement to bolus dosing for immediate need for surgical anesthesia.

Additionally, this patient presented as a twin gestation further complicating her care. Upon review of literature, there were no other reported cases of ACM-1 with twin gestations or a combined cesarean section and forceps-assisted vaginal delivery. Cesarean section delivery is needed in 10% of cases of twin B after a vaginal delivery of twin A. Malpresentation, non-reassuring FHR tracing, cephalopelvic disproportion, and cord prolapse are all predictors of emergency cesarean section for delivery of twin B7. Patients with multiple gestations should deliver in a room where cesarean section is immediately possible and 3% 2-chloroprocaine should be drawn up in anticipation of an emergent cesarean section upon non-reassuring FHR tracing. The block should be optimized to a T6-8 level in anticipation of possible version maneuvers and so that surgical level and density of anesthesia may be reached quickly if needed2.

Conclusion: Chiari malformations pose a clinical dilemma for the anesthesiologist. The risk of exacerbation of cerebellar tonsillar herniation and the safety of neuraxial anesthesia have both been questioned. Current review of the literature does support the safety of neuraxial and general anesthesia in the parturient. No currently available clinical data suggests an increased risk for high epidural in these patients. This is the only case report which describes a parturient with ACM I, with twin gestation and functioning epidural, requiring an immediate conversion to cesarean section after the successful delivery of one twin via vaginal delivery with labor epidural. Based on our experience, a well-functioning epidural, if neuraxial anesthesia is utilized in an asymptomatic ACM-1, would be especially useful in patients with multiple gestations. These patients are at risk of postpartum hemorrhage and conversion to cesarean section secondary to malpresentation or non-reassuring FHR. However, current literature does not describe if parturients with a working epidural requiring emergent cesarean section should undergo general anesthesia when there is inadequate time for incremental blousing. It is our opinion, that asymptomatic ACM-1 patients with a working epidural can undergo emergent cesarean section with that epidural. Know there is a risk of increased ICP with epidural anesthesia especially with large boluses in rapid succession as is the case in an emergent cesarean section. Furthermore, as in any other case, there is a risk of high epidural and it should be identified and managed without delay.