Spinal neuraxial anaesthesia for Caesarean section in a parturient with Type I Arnold–Chiari malformation and syringomyelia

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Abstract
Type I Arnold–Chiari Malformation is associated with prolapse of the cerebellar tonsils into or below the level of the foramen magnum and is usually diagnosed in adults. There are no current guidelines for the management of patients with a residual Type I Arnold–Chiari Malformation, planned for a Caesarean section under spinal neuraxial anaesthesia. The paucity of literature on this topic presents as a management dilemma. We report a case of a term parturient with Type I Arnold–Chiari Malformation, following surgical decompression four years earlier, with a residual syringomyelia who underwent an elective Caesarean section under spinal neuraxial anaesthesia. This case highlights that multidisciplinary management and an early anaesthetic consult is of paramount importance in the outcome of the patient, and that spinal neuraxial anaesthesia can be considered as a safe anaesthetic option.

Keywords
Arnold–Chiari Malformation, pregnancy and Caesarean section, spinal neuraxial, anaesthetic management

Introduction
Arnold–Chiari Malformation (ACM) can be divided into four types (Types I–IV). Type I is a congenital neurological anomaly with prolapse of the cerebellar tonsils into or below the level of the foramen magnum and is usually diagnosed in adults. Between 30% and 50% of patients have an associated syringomyelia. They typically present with headaches, neck and shoulder pain, paraesthesia and mild incoordination. Most are asymptomatic and discovered incidentally on brain or cervical magnetic resonance imaging (MRI) scans.1–3

Suboccipital craniectomy and decompression has been of modest benefit in improving the symptoms caused by craniospinal pressure dissociation associated with the disordered flow of cerebrospinal fluid (CSF). Best results are obtained when surgery is performed relatively early during the course of the disease.

There have been case reports of the use of spinal neuraxial anaesthesia for Caesarean sections in such patients, though the information is limited with no consensus on the topic thus far. We are presenting a parturient with Type I ACM and syringomyelia planned for a Caesarean section.

Case report
A 40 year old female, height 145 cm, weight 45 kg, Gravida 1 Para 0, was planned for an elective Caesarean section at 38 weeks of gestational age. She has a significant past medical history of thalassemia minor, migraine and a Type I ACM. She initially presented five years ago with persistent left sided headaches, numbness of the left side of the face, upper limb and neck. The initial MRI depicted a large syringohydromyelia seen in the entire spinal cord extending from C1 to T12 level with only a small syrinx in the conus medullaris. There was herniation of cerebellar tonsils below the level of foramen magnum by about 11 mm. She underwent a foramen magnum decompression. Intraoperative findings were that of a

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thickened dural band at the foramen magnum, with elongation of tonsils down to the upper border of C1. The follow-up MRI showed that the CSF space around the craniocervical junction had improved, though the large syrinx still persists and extends from C1 to beyond T5.

Neurosurgical consult was made early in pregnancy by the obstetrician and she was deemed suitable to proceed with either a normal vaginal delivery or Caesarean section, and both a general or a regional anaesthetic technique was deemed appropriate.

Pre-operative assessment was conducted one day prior to the surgery. There was residual left sided numbness over her face and left upper limb and the headaches were still present but not worsening. There were no other signs or symptoms of raised intracranial pressure. These symptoms were not aggravated with pregnancy, coughing or neck movements.

A physical examination was done; there was decreased sensation over the left face and upper limb. Airway examination was unremarkable. There was a slight thoracic scoliosis over her back but the spinous processes were clearly felt in the lumbar region.

She was counselled regarding the risks and benefits of spinal (neuraxial anaesthesia) versus general anaesthesia. A normal vaginal delivery was discussed with the patient again as it was not a contraindication. However, she had concerns about lying supine for a prolonged period due to her thoracic scoliosis.

She had initially decided to proceed with the Caesarean section under a general anaesthetic. However, after thorough discussion of the risks and benefits of a general versus a spinal neuraxial anaesthesia technique, the decision was made to proceed with the surgery under a spinal neuraxial anaesthesia.

The patient was positioned in the sitting position, a single shot spinal anaesthetic was given at the level of L3/4 interspace. This was a single attempt using a 27 gauge Whitacre spinal needle, with a spinal introducer. Given her relatively short stature, intra-thecal hyperbaric bupivacaine 0.5% 1.6 ml with fentanyl 15 µg and morphine 100 µg was given. Assessment of block height to cold sensation was performed, and a level corresponding to that of T4 was achieved prior to surgical incision.

Intra-operatively, the patient had mild discomfort from intraoperative surgical stretching of the abdomen, which was transient in nature. She was given ivaneous fentanyl 50 µg in titrated boluses, together with entonox prior to the delivery of the baby. After the delivery of the baby, she was given intravenously 1 mg of midazolam. The surgery was completed uneventfully without further complaints. There was no worsening of symptoms intra-operatively after the single shot spinal anaesthetic. Post-operatively, she was monitored in the high dependency unit overnight. Follow up on the patient on the first day following surgery revealed no worsening of her pre-existing neurological symptoms, which was that of a left sided numbness over her face and left upper limb. There was also no interval development of new neurological symptoms. She was discharged on post-operative day 3 and remained well on her follow-up visit with the obstetrician two weeks later.

**Discussion**

There is a lack of literature on the management of a pregnancy in a woman affected by ACM. There is little information regarding the safety of allowing labour or the use of neuraxial anaesthesia in women with corrected ACM, and these anaesthetic practices have been restricted to case reports. An ACM with a concomitant syringomyelia implies that there is an initial or persisting continuity between the syrinx and CSF in the central canal of the cord. The foramen magnum abnormalities cause intermittent obstruction to CSF outflow from the fourth ventricle, with development of craniospinal pressure dissociation with a relatively higher CSF pressure in the head and a lower pressure in the spine. In patients with an uncorrected ACM, any disorder of the posterior fossa should be a contraindication to lumbar puncture as this might cause herniation of the cerebellar tonsils. Traditionally, anaesthetic management of these patients would be general anaesthesia, avoidance of CSF pressure fluctuation and intracranial pressure (ICP) elevations. However, risks and benefits of each technique should be weighed before proceeding.

This case highlights the importance of an early multidisciplinary approach to a complex issue with an individualised plan for the patient. It is commonly perceived that once surgical decompression has been carried out, there will be no further concerns. However, some patients may have residual disease and the use of standard neuraxial analgesic and anaesthetic techniques without any knowledge of the ongoing CSF pressure may in fact be harmful. As most anaesthesiologists do not have the background or training to evaluate an MRI, it is important to evaluate the scans with a neurosurgeon/radiologist, with emphasis on the cerebellar tonsils and cervical cord, prior to anaesthetic selection.

In this case, the patient had opted for an elective Caesarean section. She was offered a spinal neuraxia anaesthesia. There are theoretical concerns with spinal neuraxial anaesthesia in uncorrected or symptomatic patients with Type I ACM because of the potential for compression of structures at the level of the foramen magnum, increased ICP or obstructive hydrocephalus, and intramedullary cervical cord syndrome. There have been reports of worsening of symptoms up to two weeks after a spinal anaesthetic was administered. In two case reports, the ACM was unknown at the time of delivery and only diagnosed subsequently because of adverse neurological outcomes. There have also been reports of successful spinal neuraxial anaesthesia in women with surgically corrected Type I ACM. In these cases, there was active involvement of the neurosurgical team, and it was determined that a neuraxial anaesthesia was not contraindicated because a dural puncture should neither impair CSF flux nor precipitate bulbcompression.

We opted for a single shot spinal anaesthetic with a 27G Whitacre needle as our anaesthetic of choice. This would reduce the risk of local anaesthetic toxicity and avoid the risk of an inadvertent dural tap with an 18G Tuohy needle when attempting an epidural anaesthetic. A single shot spinal anaesthetic with hyperbaric bupivacaine, fentanyl and/or morphine may be more beneficial in the detection of neurologic disturbances when compared with an epidural which requires...
repeated dosing. Epidural anaesthesia might raise the ICP when a bolus of local anaesthetic is given due to dural compression in the epidural space with a shift of CSF into the cranium. However, slow and small increments of local anaesthetic in the epidural space may circumvent this issue. A neuraxial technique may offer several advantages in that it avoids the potential hazards of securing the airway, respiratory function is less compromised and the existing craniospinal CSF pressure relationship is better preserved.

When considering a neuraxial technique, it is important to look for associated abnormalities such as kyphoscoliosis and spina bifida. Neurological assessment should include a check for autonomic neuropathy. Its identification is paramount as these patients commonly present with tachyarrhythmias and widely fluctuating arterial pressures in response to stress and anaesthesia. Sudden cardiac and respiratory arrests have also been attributed to autonomic neuropathy in patients with syringomyelia after posterior fossa surgery.

GA requires careful prior airway assessment and planning. Intubation may be complicated by the need to avoid succinylcholine if significant muscle denervation has occurred, due to hyperkalaemia and transient increase in ICP from fasciculations. This should be weighed against the risk of aspiration in parturient and delayed gastric emptying, which is also associated with ACM. Patients may be sensitive to non-depolarizing neuromuscular blockers due to muscle wasting and atrophy. Specific attempts should be made to control the increase in ICP and to avoid CSF pressure fluctuations during intubation and extubation.

Conclusion
Multidisciplinary management between the anaesthetic, obstetric and neurosurgical teams should be established before conception. Patients with a surgically corrected ACM should have a MRI post surgery. If the MRI is not available, efforts to engage in a consultation with the neurosurgeon is necessary even if they are asymptomatic. There should be an individualized approach to parturients with uncommon, non-pregnancy specific disorders such as Type 1 ACM. This case highlights that a spinal neuraxial anaesthesia is a safe and effective option in parturients with a corrected ACM and syringomyelia undergoing a Caesarean section, given that careful planning has been undertaken.

Conflict of interest
The author certifies that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers’ bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

Funding
This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

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