

Successful Epidural Placement and Vaginal Delivery In a Case of Arnold-Chiari Malformation type 1 and Ehler Danlos Syndrome with the Assistance of Point of Care Ultrasound and Epidural Needle Stopper

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Introduction

The association between Arnold-Chiari Malformation type1 (ACM-1)and Ehler's Danlos Syndrome(EDS) has been described recently¹. The anesthetic management of parturients needing labor anesthesia or anesthesia for cesarean section is a subject of discussion. When both diseases occur simultaneously, the decision is challenging and more complex.

1.Henderson, F. et al. Neurological and spinal manifestations of the Ehlers-Danlos syndromes. American Journal of Medical Genetics Part C (Seminars in Medical Genetics) 2017; 175C:195–211.

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Case Presentation

- 25-year-old G1P0 at 38 weeks of gestation with ACM1, EDS, POTS, and scoliosis.
- The diagnosis of ACM-1 was made at the age of 17 when brain imaging was performed for recurrent headaches. The patient had been symptomatic since and had been followed by the neurosurgery team who wanted to perform decompression but was postponed.
 - During the anesthesia evaluation, the patient was suffering from **persistent occipital headaches** and worsening on Valsalva which warranted repeat imaging.
 - Brain MRI : ACM-1 with **0.7 cm transtonsilar herniation** below the foramen magnum.
 - POCUS: baseline **ocular ultrasound** showed normal optic sheath diameters, no retained food on baseline **gastric ultrasound** after a small meal 4 hours previously, and the **neuraxial US** showing scoliosis with depths measured.
 - Maternal-fetal Medicine specialist, neurosurgeon, neuro-ophthalmology, and obstetric anesthesiology specialists agreed on proceeding with normal vaginal delivery and epidural for labor analgesia.

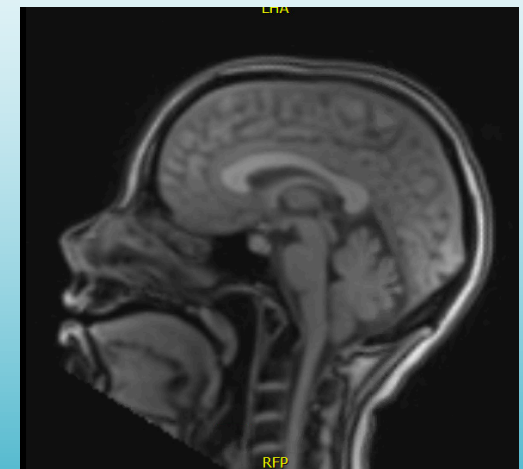


Figure 1: Brain MRI showing ACM-1 with 0.7 cm transtonsilar herniation below the foramen magnum



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Case

- The patient presented in labor requesting her epidural without any new neurologic changes. Due to POTS, she was placed in the right lateral position due to previous US showing deeper right articular process with repeat ultrasound locating L2-3 space with measurement showing depth to place epidural stopper and angle of needle trajectory noted.
- Successful lumbar epidural after one attempt for labor analgesia. The patient went on to have a normal vaginal delivery.

Discussion

- The safety of neuraxial anesthesia in patients with ACM1 was described before and showed to be safe especially when patient is asymptomatic².
- However, concerns about increased ICP and herniation are present when managing a parturient case with ACM1 and connective tissue disorder at the same time among other comorbid disorders such as POTS, GI motility disorders, and scoliosis.
- The use of POCUS can be a powerful tool to take the best care of these patients.
- The use of neuraxial ultrasound can be especially beneficial in finding exact measurements to find the epidural space and prevent any possible catastrophic side effects.



Figure 2: A-Transverse View-Neuroaxial ultrasound -B- epidural stopper added to the epidural needle