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Commentary

Chiari I malformation and delivery

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Since 1995, we have been a part of two multidisciplinary centers focused on the diagnosis and treatment of Chiari I malformation (CMI). Because of the 3:1 female prevalence of CMI, and the fact that a large number of patients become symptomatic during their reproductive years, managing a CMI patient during pregnancy and delivery is not an uncommon scenario.

The following recommendations have been adopted and field tested on the patients of our centers for more than 10 years. They reflect the feedback from experts in the field of Obstetrics and Anesthesia, as well as the evolution of our understanding of CMI and syringomyelia.

- A trial of natural childbirth is not contraindicated in patients with CMI (untreated, or after surgical decompression)
- Epidural anesthesia, if necessary, should be performed with caution to avoid dural puncture (est. 1–2% incidence), which could potentially aggravate Chiari symptoms resulting from cerebrospinal fluid (CSF) leakage. In case of dural puncture, a blood patch should be performed immediately
- Spinal anesthesia is not contraindicated, although the potential for anesthetic-related arachnoiditis is low, but unpredictable. In case of symptomatic CSF leak, a blood patch should be performed immediately
- If vaginal delivery is anticipated to be difficult, prolonged, or complicated, the threshold for a C-section with general or spinal anesthetic should be lower in undecompressed and partially decompressed patients with Chiari I, when compared to that of the normal population
- For general anesthesia, extreme and prolonged neck extension during intubation must be avoided
- Anesthesiologists should refrain from the use of anesthetic agents, such as fentanyl and ketamine, that may increase intracranial pressure
- If a very large syringomyelia cavity is present at the beginning of the pregnancy, magnetic resonance imaging (MRI) should be obtained on the 25th week of gestation, and a follow-up with the treating neurosurgeon should follow. If the syringomyelia has become larger and/or if the patient has developed a new or worsening neurological deficit, consideration should be given for an early delivery at that point (25th week or later) followed by Chiari decompression. The same criteria will apply to syringobulbia
- The most important dimension of a syringomyelia cavity is the cross diameter, not its length. A central Syrinx is problematic when the cross diameter is equal or more than 75% of the cross diameter of the spinal cord at that level. An exception to this rule is the case in which the Syrinx is asymmetric, with a side bleb rupturing in the cord parenchyma; this scenario invariably leads to a neurological deficit and represents an indication for urgent surgery regardless of its diameter
- The possible presence of recognized CMI comorbidities (such as Ehlers–Danlos syndrome, craniocervical instability, Klippel–Feil anomaly, tethered cord, mast cell activation disorder, dysautonomia, hormonal imbalance, postural orthostatic tachycardia syndrome, idiopathic intracranial hypertension, intracranial hypotension, etc.) should be kept in mind when managing CMI patients in a delivery setting.

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