

## Epidural anesthesia for labor pain and cesarean section in a parturient with arthrogryposis multiplex congenita

Sir,

A 28-year-old, G1P0, 37-week pregnant, African-American woman was admitted to the labor unit for elective trial of labor. She was 149 cm tall and weighed 68 kg with the body mass index of 30.3. She had a history of severe arthrogryposis, diabetes, gastroesophageal reflux disease, and seizures. The physical examination showed a gravid patient with severe contractures of all extremities and kyphoscoliosis of the thoracolumbar spine. Examination of the back revealed the absence of usual bony landmarks. She had Mallampati Class I airway. The vital signs and laboratory values were unremarkable. The chest X-ray showed severe thoracic kyphosis with a marked decrease in the left lung volume. We requested flexion–extension X-ray of the cervical spine, which turned out to be normal.

Induction of labor was conducted electively. Peripheral intravenous access was unsuccessful. Subsequently, a triple lumen central venous catheter was placed. After 8 h of induction of labor, we were consulted again for the management of labor pain. L4–L5 space was identified approximately. An 18-gauge Tuohy needle was used, and epidural space was identified by loss of resistance at the depth of 8 cm with lot of difficulties. A 26-gauge Gertie Marx spinal needle was then inserted through the Tuohy needle. After the confirmation of clear flow of cerebrospinal fluid (CSF), 1 cc of 0.2% plain bupivacaine was injected. Subsequently, a

20-gauge epidural catheter was inserted and taped at the skin at 14 cm. Both the spinal and epidural components worked well with sensory block up to T8 bilaterally. After another 12 h of labor, it was decided to perform cesarean section secondary to the failure of progression of labor. Epidural was slowly bolused with 20 cc of 2% lidocaine with epinephrine, and a level of T4 was achieved bilaterally. Surgery was allowed to proceed, and a healthy baby delivered uneventfully. The epidural worked successfully throughout the procedure.

Many of these patients may have facial abnormalities such as microcephaly, mandibular hypoplasia, craniosynostosis, cleft palate, and tracheal or laryngeal stenosis, making the airway management potentially challenging.<sup>[1]</sup> The other major concern is the abnormalities of the cervical spine. If it is not recognized before the surgery, forward subluxation of the spine may lead to quadriplegia.<sup>[2]</sup> These patients should have flexion–extension X-ray of the cervical spine preoperatively to rule out that possibility.<sup>[2]</sup> In addition, patients may have abnormal CSF dynamics with an unpredictable action of local anesthetics due to abnormalities such as spina bifida and sacral agenesis.<sup>[2,3]</sup> In the presence of tethered cord syndrome, these patients may also have neuraxial blockade which may be associated with serious complications such as hematoma, dural puncture, and nerve damage.<sup>[4,5]</sup> Baines *et al.* did not find any report of malignant hyperthermia in their review of 67 patients of this disease despite receiving 398 anesthetics.<sup>[6]</sup> These patients can have a hypermetabolic response with hyperthermia even without exposure to triggering agents as in the cases of study by Hopkins *et al.*<sup>[7]</sup> It can be treated with active cooling, but dantrolene is not indicated. Cardiac anomalies may also be associated with this disorder, but without any definitive pattern.<sup>[1]</sup>

Our patient came to the labor floor for elective trial of labor. In spite of the known structural abnormalities of the spine, advantages of the regional anesthesia outweighed the risks of general anesthesia. The major reason for our choice of combined spinal epidural was that the patient wanted a trial of labor, which allowed us additional time to test the quality of block. Second, the spinal component of combined spinal and epidural anesthesia and flow of CSF provides an additional confirmation about the position of the epidural needle as the loss of resistance can be subtle in these patients. Finally, we wanted to avoid postdural puncture headache (PDPH) in this patient secondary to the placement of an elective intrathecal catheter, since treatment of PDPH would have been very difficult in the postpartum period.

Epidural anesthesia is an option for the treatment of labor pain in these patients. Although never tried in the past, the successful use of combined spinal and epidural anesthesia in our case suggests that it can be a viable epidural anesthesia for labor pain and cesarean section in a parturient with arthrogryposis multiplex congenita as an option for the treatment of labor pain

in this population. We suggest that these patients should be assessed early by the anesthesiologists to identify the relevant comorbidities; hence, thorough planning should be made for a safe conduct of anesthesia for labor and delivery.

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### Conflicts of interest

There are no conflicts of interest.

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### References

1. Martin S, Tobias JD. Perioperative care of the child with arthrogryposis. *Paediatr Anaesth* 2006;16:31-7.
2. Steinberg B, Nelson VS, Feinberg SE, Calhoun C. Incidence of maxillofacial involvement in arthrogryposis multiplex congenita. *J Oral Maxillofac Surg* 1996;54:956-9.
3. Quance DR. Anaesthetic management of an obstetrical patient with arthrogryposis multiplex congenita. *Can J Anaesth* 1988;35:612-4.
4. Wood GG, Jacka MJ. Spinal hematoma following spinal anesthesia in a patient with spina bifida occulta. *Anesthesiology* 1997;87:983-4.
5. Tidmarsh MD, May AE. Epidural anaesthesia and neural tube defects. *Int J Obstet Anesth* 1998;7:111-4.
6. Baines DB, Douglas ID, Overton JH. Anaesthesia for patients with arthrogryposis multiplex congenita: What is the risk of malignant hyperthermia? *Anaesth Intensive Care* 1986;14:370-2.
7. Hopkins PM, Ellis FR, Halsall PJ. Hypermetabolism in arthrogryposis multiplex congenita. *Anaesthesia* 1991;46:374-5.

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