The Journal of Maternal-Fetal & Neonatal Medicine

ISSN: 1476-7058 (Print) 1476-4954 (Online) Journal homepage: informahealthcare.com/journals/ijmf20

Combined spinal epidural anesthesia for cesarean section in a parturient with spinal muscle atrophy type III (Kugelberg–Walendar disease)

Michal Gaca, Natalia Kokot, Agnieszka Koziolek & Krzysztof M. Kuczkowski

To cite this article: Michal Gaca, Natalia Kokot, Agnieszka Koziolek & Krzysztof M. Kuczkowski (2011) Combined spinal epidural anesthesia for cesarean section in a parturient with spinal muscle atrophy type III (Kugelberg–Walendar disease), The Journal of Maternal-Fetal & Neonatal Medicine, 24:1, 195-195, DOI: 10.3109/14767058.2010.482617

To link to this article: https://doi.org/10.3109/14767058.2010.482617

Published online: 23 Jun 2010.

The Journal of Maternal-Fetal and Neonatal Medicine, January 2011; 24(1): 195

informa
healthcare

LETTER TO THE EDITOR

Combined spinal epidural anesthesia for cesarean section in a parturient with spinal muscle atrophy type III (Kugelberg–Walendar disease)

MICHAL GACA[1], NATALIA KOKOT[1], AGNIESZKA KOZIOLEK[1], & KRZYSZTOF M. KUCZKOWSKI[2]

[1]*Department of Anaesthesiology and Intensive Therapy, Karol Marcinkowski Medical University, Poznan, Poland and* [2]*Department of Obstetrics and Gynecology, University Medical Center of El Paso, El Paso, Texas, USA*

Spinal muscular atrophy (SMA) is an autosomal recessive neurodegenerative disease characterized by the degeneration of spinal cord motor neurons, atrophy of skeletal muscles, and generalized weakness [1]. The use of central neuraxial blockade in patients with neurodegenerative disease and preexisting neurological disorders is still controversial (theoretical risks of worsening of neurological symptoms following regional anesthesia). We herein report a case of a parturient with SMA disease who received an uneventful combined spinal epidural anesthesia (CSEA) for cesarean section.

A 32-year-old G1P0 female at 37 weeks of gestation and coexisting SMA disease type III (Kugelberg–Welander disease) required elective cesarean section (SMA disease and fetal breech presentation). Grade 4 muscle weakness mainly affecting her lower extremities was noted on preanesthetic neurological examination. A standard needle-through-needle CSEA technique (at the L2–L3 interspace) with subarachnoid injection of 14 mg of 0.5% (heavy) bupivacaine induction dose was selected for the surgery. A T-5 dermatome sensory level of anesthesia was established within 9 min of induction of anesthesia. The motor block was difficult to evaluate secondary to the preexisting SMA-related neurological muscle weakness. The surgery was uneventful and patient's motor function (lower extremities) returned to preanesthetic baseline within 7 h of spinal dose. No worsening of SMA disease was noted in the postoperative period and at follow up visit 2 weeks after delivery.

SMA is an autosomal recessive neuromuscular disorder characterized by muscle weakness and atrophy due to degeneration of motor neurons of the spinal cord and cranial motor nuclei [1]. The SMA disease is caused by homozygous disruption of the survival motor neuron 1 (SMN1) gene by deletion, conversion or mutation and results in insufficient levels of SMN protein in motor neurons [2]. The clinical phenotype incorporates a wide spectrum. No effective treatment is currently available and patients may experience severe physical disability which is often life limiting. The International Standard of Care Committee for SMA was formed in 2005, with a goal of establishing practice guidelines for clinical care of these patients [3]. SMA in pregnancy is very rare and poses multiple problems for the peripartum care team [4]. We found a handful of previously published reports of regional anesthesia for cesarean section in this condition in the English language literature [5]. Our report provides one more piece of evidence that regional anesthesia is a safe alternative to general anesthesia in parturients with SMA disease.

It is concluded that evidence-based practice standards for SMA are urgently needed to help with the multidisciplinary care (including obstetric and obstetric anesthesia) of these patients.

References

1. Prior TW. Perspectives and diagnostic considerations in spinal muscular atrophy. Genet Med 2010;Jan 6. [Epub ahead of print].
2. Farrar MA, Johnston HM, Grattan-Smith P, Turner A, Kiernan MC. Spinal muscular atrophy: molecular mechanisms. Curr Mol Med 2009;9:851–862.
3. Wang CH, Finkel RS, Bertini ES, Schroth M, Simonds A, Wong B, Aloysius A, Morrison L, Main M, Crawford TO, et al. Participants of the International Conference on SMA Standard of Care. Consensus statement for standard of care in spinal muscular atrophy. J Child Neurol 2007;22:1027–1049.
4. Flunt D, Andreadis N, Menadue C, Welsh AW. Clinical commentary: obstetric and respiratory management of pregnancy with severe spinal muscular atrophy. Obstet Gynecol Int 2009;94:23–25.
5. McLoughlin L, Bhagvat P. Anaesthesia for caesarean section in spinal muscular atrophy type III. Int J Obstet Anesth 2004;13:192–195.

Correspondence: Krzysztof M. Kuczkowski, MD, Department of Anesthesiology, University Medical Center of El Paso, 4800 Alberta Avenue, El Paso, Texas, USA. Tel: +1-858-531-6400. Fax: +1-858-638-8168. E-mail: kmkuczkowski@gmail.com

ISSN 1476-7058 print/ISSN 1476-4954 online
DOI: 10.3109/14767058.2010.482617