Combined Spinal Epidural Anesthesia For Cesarean Section In A Parturient With Spinal Muscle Atrophy Type III (Kugelberg-Walendar Disease)

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Introduction: Spinal muscular atrophy (SMA) is an autosomal recessive neurodegenerative disease characterized by 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. We herein report a case of a parturient with SMA disease who received an uneventful combined spinal epidural anesthesia (CSEA) for cesarean section.

Report of case: 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 minutes 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 hours of spinal dose. No worsening of SMA disease was noted in the postoperative period and at follow up visit 2 weeks after delivery.

Discussion: 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. 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. Spinal muscular atrophy in pregnancy is very rare and poses multiple problems for the peripartum care team (including an anesthesiologist). We found a handful of previously published reports of regional anesthesia for cesarean section in this condition in the English language literature. Our report provides one more piece of evidence that regional anesthesia is a safe alternative to general anesthesia in parturients with SMA disease.

Conclusion: An evidence based practice standards for SMA are urgently needed to help with the multidisciplinary care (including obstetric anesthesia) of these patients.