Anaesthesia for Cesarean section in a patient with Huntington's Disease

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Introduction: Huntington's Disease (HD) is a premature neurodegenerative disorder with an incidence of 4-10/100,000 in the US. Symptoms appear between 30 and 45 years of age and include personality problems, dementia and choreiform movements. Juvenile HD manifests before age of 20. We report our experience of spinal anesthesia for a parturient with HD undergoing cesarean section.

Case report: A 20-year-old primipara was booked for elective LSCS. She was diagnosed with HD while eight weeks pregnant. Symptoms prior to pregnancy were thought to be due to illicit drug abuse. Her symptoms were occasional twitching, memory loss, tingling sensations in the upper limbs, eye rolling, restless legs, insomnia and erratic behavior. Her booking weight and height were 61.1 kg and 1.51m respectively. Airway and spine assessment were unremarkable. Spinal anaesthetic was administered with 2.2ml of 0.5% heavy Bupivacaine and 300 micrograms of diamorphine. A live baby girl was delivered and the procedure went uneventful albeit involuntary movement of upper limbs. Estimated blood loss during peripartum was 900 ml, managed with fluids and uterotonic support.

Discussion: Experience with anesthetic management of HD is limited to very few case reports in the nonobstetric population. These reports suggest that they are very sensitive to anesthetic agents and are at increased risk of pulmonary aspiration due to altered airway reflex (1). Generalized spasms triggered by pain and shivering can make labor difficult. Delayed awakening with thiopentone induction and prolonged paralysis with suxamethonium were initially reported in 1966. Since then, they have been used with caution in patients with HD. Propofol and Remifentanil TIVA has also been used to facilitate rapid recovery. Despite being used in HD patients, Isoflurane accelerates apoptosis in the neurons made vulnerable by the disease by an unknown mechanism (HDlighthouse.org). All NDMRs have been safely administered. Metoclopramide can exacerbate choreiform movements.

There are two successful case reports regarding use of spinal anesthetic in patients with HD (2). In severe cases of HD, positioning for spinal anesthetic can be challenging and intraoperatively involuntary movements of upper part of the body can be troublesome for monitoring and also to keep the patient still.

Conclusion: Anesthetic management of parturients with HD can be challenging. OB anesthesiologists should be aware of increased sensitivity to anesthetic drugs and increased risks of aspiration in this group of patients should a GA be required. We found spinal anesthetic to be a safe option in minimally symptomatic patient without untoward effect to mother and the baby.