Anesthesia for Labor and Delivery in a Patient with Isaacs Syndrome

Presenting Author: Patricia L Dalby Assistant Professor
Presenting Author's Institution: University of Pittsburgh - Pittsburgh, PA
Co-Authors: Guarev Rajpal MD - University of Pittsburgh - Pittsburgh, PA
Joel Pomerantz MD - University of Pittsburgh - Pittsburgh, PA
Jonathan Waters MD - University of Pittsburgh - Pittsburgh, PA
Ryan Romeo MD - University of Pittsburgh - Pittsburgh, PA
Manuel Vallejo MD - University of Pittsburgh - Pittsburgh, PA

Abstract Body: Isaacs Syndrome (IS) is a rare peripheral motor neuron disorder. It is also known as neuromyotonia, continuous muscle fiber activity, and quantal squander syndrome. Characterized by hyperexcitability of peripheral nerve axons that activate skeletal muscle fibers, it can be hereditary or acquired. The onset is in the same age as childbearing for women.

Our patient was a 22 yo 35,5/7 week primiparous female with a healthy gestation, who presented with pain and premature rupture of membranes to the emergency room. She was admitted for labor induction. Her past medical history included a bicornuate uterus, a "seizure d/o" of unknown etiology and GERD. Past surgical history included removal of lymph nodes with local infiltration and no previous general anesthesia. Her medications were ranitidine and carbamazepine, with NKDA. Symptoms of the IS included involuntary fine muscle contractions of her trunk and extremity areas, with associated stiffness and pain. She occasionally staggered, but had no involvement of her cranial-facial or laryngeal muscles. She had a prolonged induction of labor with IV oxytocin and misoprostol vaginal suppositories, for 32 hours, receiving a labor epidural at 39 hours. Epidural placement and lidocaine test dose were uneventful, and she enjoyed a PCEA infusion of bupivacaine 0.08% with fentanyl 2mcg/cc at 8 cc/hr CEI, an optional 8 cc bolus of CEI every 8 minutes, and a possible 24 cc CEI per hour. Oral medication with carbamazepine was continued. She delivered a healthy male baby vaginally after 51 hours of labor. Placental pathology indicated chorioamnionitis. Baby and she had an uneventful postpartum course.

Continuous fine skeletal muscle vibrations (myokymia) with diagnostic EMG findings of IS, may be associated with symptoms of ataxia, and symptoms affecting bulbar muscles. Progressive muscle stiffness may develop with delayed muscle relaxation and associated muscle pain. Increased sweating has occurred. Acquired IS can develop in association with paraneoplastic syndromes, peripheral neuropathies, and after radiation therapy, but most commonly in conjunction with the autoimmune illnesses such as thymoma, Hashimoto's thyroiditis, and myasthenia gravis. The autoimmune form has antibodies that bind to potassium channels on the motor nerves. Treatment includes phenytoin, acetazolamide and carbamazepine, and IV immunoglobulin.

Safe administration of labor epidural anesthesia to a parturient with IS has been previously reported once.(1) Muscle fasciculations have been seen to be persistent even after general anesthesia, while succinylcholine and peri-dural regional anesthesia attenuate them. A recent report was written about exquisite sensitivity of a patient with IS to Rocuronium.(2) Bulbar muscle weakness in IS has increased aspiration risk. “Stiff man syndrome” has been reported with IS and associated with difficult ventilation and intubation.

(2)Muscle Nerve 40:p139, 2009