Cesarean section in a patient with Pena-Shokeir syndrome, with severe scoliosis despite Harrington rod correction and known malignant hyperthermia history

Presenting Author: Izabella Wasiluk MD
Presenting Author's Institution: University of Florida, College of Medicine, Jacksonville - Jacksonville, FL
Co-Authors: Natesan Manimekalai MD - University of Florida, College of Medicine, Jacksonville - Jacksonville, FL
Moeen Panni MD PhD - University of Florida, College of Medicine, Jacksonville - Jacksonville, FL

Introduction: Regional anesthesia is challenging in patients with spinal scoliosis, especially those with Harrington rods. We report a case of a 36 week gestation parturient who presented for a planned C-section with Pena-Shokeir syndrome, severe spinal scoliosis, severe myopathy, cleft palate surgical repair, history of malignant hyperthermia (MH)/prolonged intubation and tracheostomy placement.

Case: 21 year old female, G1P0 with Pena-Shokeir syndrome with severe myopathy and extreme weakness in her lower extremities, presented to us for a planned C-section. She had contractures of all her major joints, a cleft palate and severe congenital scoliosis limiting her mobility (wheelchair bound). PSH included cleft palate repair, G tube placement, correction of scoliosis with rod placement between T1-L5 level, with post operative severe respiratory failure (tracheotomy placed) following an episode of MH after her first surgery. The patient was of short stature (1.3m, 35.5 Kg) Mallampati Class II airway with a large tongue and protruding upper incisors and limited C-spine extension and neck tracheostomy scar. Palpation of her spine did not reveal any useful landmarks except the scar extending from the lower cervical to the lower lumbar region. At 36 weeks pregnancy, she was initially admitted with sudden onset of dyspnea, tachypnea, palpitation, and chest pain. Chest CT ruled out pulmonary embolism. This resolved and the plan was to proceed to C-section the following day. She received a single shot spinal anesthetic (7.5mg bupivacaine) at the L5-S1 level (below the scar termination) in the sitting position, with a plan to convert to a non-triggering general anesthetic with MH precautions (with difficult airway equipment present). The patient was then placed supine with left uterine displacement. After 10 minutes, there was T10 level of surgical anesthetic block and by 12 minutes, T6 level was reached. C-section was then performed and a live female baby, 5lbs and 8oz, 9-9 apgar scores, was delivered, normal in appearance without any deformities noted. The surgery lasted 70 minutes and no supplemental medications were needed for anesthesia. Post operative period was uneventful and patient was discharged four days later in good condition.

Discussion: Pena-Shokeir syndrome is an inherited disorder characterized by arthrogryposis, facial anomalies, pulmonary hypoplasia and dysmorphic features resulting from fetal akinesia. This is the first report of a patient with Pena-Shokeir syndrome who survived up to 21yrs old and delivered a normal baby by C-section. Appropriate preparations were made for the use of a non-triggering general anesthetic, a short stature patient, with known myopathy/history of MH and prior tracheostomy, with difficult airway equipment available, however she received an uncomplicated single shot spinal bupivacaine anesthetic successfully, despite the short stature and potential challenges of spinal anesthetic spread.