Cesarean Section in an Obstetric Patient with Pena-Shokeir Syndrome Type-1, with Partially Corrected Scoliosis with Spinal Rod Placement and History of Malignant Hyperthermia

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Abstract

We report a patient with Pena-Shokeir syndrome with severe scoliosis corrected with spinal rod placement from T1-L5, with a known history of malignant hyperthermia presenting for cesarean section. A 21 year old female, 132 cm tall and 35.5 Kg presenting for a primary C-section at 38 weeks with sudden onset of dyspnea, tachypnea, palpitation, and chest pain. A spiral CT demonstrated no pulmonary embolus or any other pathology and clinical symptoms resolved with oxygen and albuterol therapy. The patient had a history of Pena-Shokeir syndrome which included severe myopathy and extreme lower extremity weakness since birth. The patient had contractures of all her major joints, involving both the upper and lower extremities. Additionally she also had cleft palate and severe congenital scoliosis significantly limiting her mobility. Past surgical history included cleft palate repair in childhood, and correction of scoliosis with spinal rod placement between T1- L5 vertebral levels, 7 years earlier, with a very difficult peri-operative course complicated by malignant hyperthermia intra-operatively and severe respiratory failure requiring prolonged mechanical ventilation and tracheostomy formation. A spinal anesthetic was administered (7.5mg hyperbaric bupivacaine only) at the L5-S1 level on the first attempt with positive CSF. A surgical anesthetic level (T6) was obtained and an uncomplicated C-section was performed. This is the first case report of a parturient with Pena-Shokeir syndrome surviving to 21 years of age and then delivering a normal baby by Caesarean section. Despite numerous anesthetic and medical concerns, she received a successful spinal anesthetic and had an uneventful delivery. Early consultation with obstetric and anesthesia team during the antepartum period aided in developing an appropriate clinical plan for this patient.

Keywords: Pena-Shokeir syndrome; Malignant hyperthermia; Scoliosis

Introduction

Regional anesthesia is the preferred method for operative delivery in parturients, however certain conditions can make its’ utilization challenging. Regional anesthesia is particularly difficult in patients with surgically corrected scoliosis with spinal rod placement, usually having an increased risk for both complications as well as failure to obtain satisfactory analgesia [1]. The benefits of regional anesthesia are well recognized and include reduced mortality [2], reduced blood loss [3], the avoidance of airway instrumentation and inherent risks associated with that in pregnancy and a reduced incidence of thrombo-embolism [4]. The avoidance of general anesthesia (with triggering agents) is crucial in patients with a known history of malignant hyperthermia. We report a patient with Pena-Shokeir syndrome [5,6] with severe scoliosis corrected with spinal rod placement from T1-L5, with a known history of malignant hyperthermia presenting for elective C-section, who had an uncomplicated C-section under spinal anesthesia. Her past surgical history consisted of a previous cleft palate repair in childhood, and correction of scoliosis with spinal rod placement between T1-L5 vertebral levels, 7 year earlier, this latter surgery being associated with malignant hyperthermia and prolonged mechanical ventilation postoperatively requiring tracheostomy.

On initial preoperative presentation, the patient was of short stature (132 cm) weighed 35.5 kg and normal vital signs were recorded. Ultrasound performed at 18 weeks of gestation revealed a normal fetus with good fetal movements in utero. Airway assessment was a Mallampati score of II with a large tongue and protruding upper incisors as well as limited C-spine extension. A tracheostomy scar was evident on her neck. Palpation of her spine did not reveal any useful landmarks, with a scar that extended from lower cervical to lower lumbar region, however the level of the spinal rods was confirmed by old plain film reports. A number of clinical problems relative to anesthetic management were apparent following this preoperative evaluation. These included a positive history of malignant hyperthermia, the presence of a congenital myopathy, a potentially difficult airway and a prolonged peri-operative course complicated by malignant hyperthermia and severe respiratory failure requiring prolonged mechanical ventilation postoperatively requiring tracheostomy.

Case Report

A 21 year old female, gravida 1 para 0, 33 weeks gestation was referred by her obstetrician for anesthetic assessment for an elective C-section scheduled at 38 weeks gestation. The patient had a history of Pena-Shokeir syndrome which included severe myopathy and extreme lower extremity weakness since birth with contractures of all major joints. She did however have no developmental cognitive delay. Additionally she also had a cleft palate and severe congenital scoliosis limiting her mobility where she was mostly wheelchair bound. Muscle biopsy had been performed which had ruled out other forms of congenital myotonic muscular dystrophy and spinal muscular atrophy. 

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higher potential for failed or neuraxial technique due to the history of scoliosis surgery with rod placement. Considering the patient’s medical condition, the anesthetic plan was to attempt a single shot spinal anesthetic for a scheduled C-Section, and if unsuccessful, to proceed to total intravenous general anesthesia with malignant hyperthermia precautions using modified rapid sequence induction with rocuronium to secure the airway—clearly the avoidance of suxamethonium and inhaled anesthetic agents. Three weeks later at 38 weeks gestation, the patient was admitted with sudden onset of dyspnea, tachypnea, palpitation, and chest pain. A spiral chest CT ruled out pulmonary embolism or any other abnormality. Her clinical symptoms resolved within 2 hours with albuterol nebulizer and oxygen therapy, and given her complex clinical history (including the requirement for malignant hyperthermia precautions) and lack of fetal distress, C-section was scheduled for the subsequent morning.

Prior to surgery the anesthesia machine was purged with oxygen for one hour, with volatile agent vaporizers removed and malignant hyperthermia cart and difficult airway equipment being readily available in the operating room in anticipation of the possible administration of a general anesthetic in the event of neuraxial failure. A spinal anesthetic was administered (7.5 mg hyperbaric bupivacaine only) at the L5-S1 level on the first attempt with positive CSF and a sufficient surgical anesthetic level (T6) was achieved. An uncomplicated c-section was then performed with a Pfannenstiel incision and a live born female infant was delivered with 9 and 9 Apgar scores at 1 and 5 minutes respectively, the infant was not dysmorphic in appearance and weighed 2.27 Kg. The postoperative period was uneventful, a TEE being also performed given clinical history suggestive of pulmonary embolus at the time of presentation, which demonstrated nil of note and the patient was discharged home on the 4th post-operative day.

Discussion

Pena-Shokeir syndrome is an autosomal-recessive disorder, characterized by arthrogryposis, facial anomalies, pulmonary hypoplasia and dysmorphic features resulting from fetal akinesia [5,6]. It was first described in 1974 and was subsequently included among the phenotypes associated with fetal akinesia deformation sequence. Autosomal-recessive is the commonest pattern of inheritance. Several descriptions of unusual presentations suggest a heterogeneous etiology [7]. The actual incidence of Pena-Shokeir syndrome Type 1 is not known; approximately 80 to 100 cases have been reported in the medical literature. This syndrome occurs in both males and females and does not associate with any specific ethnic group.

Regional anesthesia can be challenging in patients with scoliosis corrected with Harrington rod placement because of the abnormal vertebral anatomy and the instrumentation from the surgery which can cause adhesions or obliteration of the epidural space [1]. This has been reported to be associated with an increased risk of complications including failure to identify the epidural space, an increased incidence of postdural puncture headache syndrome, and failed or inadequate block, all related to distorted epidural anatomy [8]. Successful use of epidural, combined spinal epidural, single shot spinal, and continuous spinal anesthesia for labor and caesarean section in patients with severe scoliosis has been described [9-11]. This patient’s fusion extended to the L5 level and given that the majority of epidural failures in patients with Harrington rods reported in the literature occurred primarily when spinal fusion extended to L3 or lower [1,12], we chose a single shot spinal technique at the L5-S1 interspace. This is the widest spinal interspace, therefore the likelihood of technical success is more likely, also the subarachnoid space is unaffected by the surgery and spread of local anesthetic is more reliable[9] when compared to the epidural space. An intrathecal catheter may also have been an option with sequential administration of local anesthetic, however its use for this specific indication was not approved at the time. Doses as low as 0.06 mg/cm height of intrathecal bupivacaine have been shown to provide effective spinal block for C-Section in normal patients [13]. Since our patient was only 4’ 4” (132 cm) tall with partially corrected scoliosis we used 7.5 mg bupivacaine (0.057 mg/cm) to maintain maternal hemodynamic stability and prevent a high spinal block which may have been deleterious.

Conclusions

This is the first case report of a parturient with Pena-Shokeir syndrome surviving to 21 years of age and then delivering a normal baby by Caesarean section. Despite numerous anesthetic and medical concerns, she received a successful spinal anesthetic and had an uneventful delivery. Early consultation with obstetric and anesthesia team during the antepartum period aided in developing an appropriate clinical plan for this patient.

References