

# Anesthetic Management for a Parturient with Facioscapulohumeral Muscular Dystrophy Undergoing Caesarean Section

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Facioscapulohumeral muscular dystrophy (FSHD) is an autosomal dominant muscle disorder characterized by progressive weakness and wasting of facial, shoulder girdle and upper arm muscles. Anesthetic management for the parturient with muscular dystrophy is very challenging for anesthesiologists because general as well as regional anesthesia may cause deleterious effect to the patient. We report a case of 28 years parturient with Facioscapulohumeral muscular dystrophy that underwent elective caesarean section under combined spinal epidural anesthesia. Intraoperative and postoperative period were uneventful however the motor block was prolonged. Regional anesthesia especially combined spinal epidural anesthesia can be safely used to provide anesthesia for caesarean section in patients with muscular dystrophy.

**Keywords:** combined spinal epidural anesthesia; caesarean section; facioscapulohumeral muscular dystrophy.

## INTRODUCTION

Facioscapulohumeral muscular dystrophy (FSHD) is an autosomal dominant muscle disorder characterized by progressive weakness and wasting of facial, shoulder girdle and upper arm muscles. It is the third most common adult muscular dystrophy with the best-estimated prevalence of 1:20000,<sup>1</sup> although a more recent estimate from Utah suggests a prevalence of 1:15000.<sup>2</sup> Anesthetic management for the parturient with muscular dystrophy is very challenging for anesthesiologists because general as well as regional anesthesia may cause deleterious effect to the patient. Here we present a successfully managed case of a parturient with FSHD who underwent elective caesarean section.

## CASE

A 28 years primigravida at 38 weeks of gestation, under regular antenatal check up, with the history of facioscapulohumeral muscular dystrophy (FSHD) was admitted to University of Malaya Medical Center, Kuala Lumpur, for elective cesarean section. Her BMI was 22.

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Her symptoms of muscle weakness started at the age of eight years but it was ignored blaming her thin built. After two years of gradual progression, it became generalized and she was diagnosed to have muscular dystrophy confirmed with muscle biopsy done under general anesthesia. At the age of 15 years, it worsened further and she developed scoliosis, which was managed with spine brace. Later she was diagnosed clinically to have FSHD. A year before, she underwent dilatation and curettage for miscarriage. She was continuing her job as a clerk until she again conceived. With the pregnancy, her symptoms further worsened and at 6 months of pregnancy she was confined to wheel chair and needed neck collar for support. At 8 months of gestation she developed gestational diabetes mellitus for which she is taking Metformin 850 mg once a day. She doesn't have any other significant medical history.

On evaluation, her pulse rate was 75 bpm, blood pressure was 120/80 mmHg, respiratory rate was 14 bpm and airway assessment with Mallampati Grade was II. Her systemic examination was within normal limit. Central nervous system examination revealed bilateral upper and lower limb weakness with power of 4/5 and minimum facial weakness. She had thoracolumbar scoliosis. Preoperative investigations revealed hemoglobin of 13.8 g/L, renal function test, and coagulation profile were within normal limits. Pulmonary function test revealed forced expiratory

volume in 1 second was 51 % and vital capacity of 46%, FEV1/FVC >100, suggestive of restrictive pattern secondary to muscular dystrophy. Echocardiography showed normal left ventricular function, ejection fraction of 73% and no other abnormalities. The chest X-ray showed normal lung fields and no cardiomegaly. Scoliosis of thoracolumbar spine was noted. Then our diagnosis was G2P0+1 at 38 weeks of pregnancy with FSHD with ASA III.

She was planned for combined spinal epidural anesthesia. Written informed consent was taken after explaining the details of the procedure and its complications. She was advised to stay nil per oral from 12 at midnight. As a part of aspiration prophylaxis, injection ranitidine 50 mg and metoclopramide 10 mg IV and sodium citrate 0.3 M 30 ml per oral was advised before entering the operation theater. General anesthesia was kept as a back-up anesthesia in case regional anesthesia failed. Thus operation theater was prepared according to the malignant hyperthermia protocol of prevention. The vaporizer was removed from the anesthetic machine and flushed with oxygen for 6 hours. Dantrolene was kept as a back-up drug for the treatment of malignant hyperthermia.

All the monitors such as noninvasive blood pressure, electrocardiography, and pulse oximeter were attached as she comfortably lied in the operation table. Her pulse rate was 85 bpm, blood pressure 140/90 mmHg and SpO<sub>2</sub> 99%. Intravenous line was secured with 18G cannula in the left hand. Under the strict aseptic precaution, with the patient in sitting position, 18 gauge Tuohy's needle was used to identify the epidural space at L3-L4 using loss of resistance technique with normal saline then 27 G spinal needle was inserted and after free flow of cerebrospinal fluid 0.5 % heavy bupivacaine 2 ml and fentanyl 15 mcg (total 2.3 ml) was given for spinal anesthesia. After that epidural catheter was fixed at 9 cm from skin level (skin to epidural space was 5 cm and 4 cm inside the epidural space). The level of block was T4. Oxygen was given via nasal prong @ 3L/min. Inj cefuroxime 1.5 gm and metronidazole 500 mg was given for prophylaxis. Except for one episode of hypotension, which was treated with phenylephrine 50 mcg, she was hemodynamically stable throughout the surgery. A healthy baby was delivered with the

APGAR score of 8/10 at 1 min and 10/10 at 5 min. Total blood loss was 250 ml. After 1 hour in recovery room, she was shifted to high dependency unit. For postoperative analgesia we started epidural infusion with 0.1% ropivacaine with 2 mcg/ml fentanyl @ 4 ml/hour.

After 7 hours of epidural infusion, she complained of pain in left side of wound. On evaluation she could lift her left leg easily but not the right leg so the epidural catheter was pulled out 1 cm. She then was kept in left lateral position and bolus dose of 5 ml ropivacaine was given, which alleviated the pain. The next morning, epidural infusion was discontinued and 4 hours later, after removal of epidural catheter, heparin 5000 IU subcutaneously was started as a thromboprophylaxis. Her motor block was fully recovered after 57 hours of surgery. She was discharged on third post-operative day after regaining her motor power.

## COMMENT

Facioscapulohumeral muscular dystrophy is a disorder characterized by muscle weakness and wasting of facial, shoulder blades and upper arms. The onset of the disease is variable, with most patients becoming symptomatic in the second decade. Both male and female are equally affected but female tends to be less severely affected than male. The progress of the disease is slow and the life expectancy is normal. It slowly affects the lower legs, hips and pelvis and may have lower back lordosis due to weak abdominal muscles and about 20% of the patients become wheel bound.<sup>3</sup> The muscle weakness progressively increases in the pregnancy due to increased demand of oxygen.

Patient may have high tone hearing loss and retinal telangiectasias but it's very rare. Cardiac involvement like atrial arrhythmia is seen in about 5% of patient and restrictive respiratory disease requiring intervention occurs in about 1% with severe muscle weakness.<sup>1</sup>

Our patient had compromised restrictive lung disease as a result of thoracolumbar scoliosis and progressive muscle weakness during pregnancy. Ciafaloni et al<sup>3</sup> in their study had shown that 24% of the pregnant women had pain and exacerbation of muscle weakness. The anesthetic consideration of FSHD is considered to be similar to other muscular dystrophies.

Both general as well as regional anesthesia may have a high risk to a patient with muscular dystrophy. So multidisciplinary team including obstetrician, anesthesiologist, neurologist as well as pulmonologist should be involved to manage these patients as we did in our case. We must assume that these patients may be malignant hyperthermia susceptibility. The operating room should be prepared according to the malignant hyperthermia protocol of prevention which include avoiding exposure to volatile agents and succinylcholine (removal of vapouriser from the anesthetic machine to avoid unintended administration), new breathing bags, filters, respiratory circuit and washed with air 10 L/min for at least 3 hours and new soda lime.<sup>4</sup> Although we performed under combined spinal epidural anesthesia, our back up anesthesia was general anesthesia so we prepared according to the protocol. Regional anesthesia should be performed whenever possible to avoid the risk of malignant hyperthermia. There are few studies that were successfully done under spinal,<sup>5</sup> combined spinal epidural<sup>6</sup> and epidural alone<sup>7</sup> in parturients with muscular dystrophy for caesarean section. Combined spinal epidural anesthesia was

chosen in our case because of rapid onset and dense block for cesarean section and to provide adequate postoperative analgesia. We were able to perform it without any intraoperative complications. But in postoperative period we found that the effect of motor block was prolonged. It took 12 hours to fully recover from epidural analgesia.

## CONCLUSIONS

The management of pregnant women with FSHD depends on the severity of the disease and is best managed by a multidisciplinary team including obstetrician, anesthesiologists, neurologist, respiratory and intensive care physicians. Regional anesthesia especially combined spinal epidural anesthesia can be safely used to provide anesthesia for caesarean section in patients with muscular dystrophy.

## DISCLOSURE

The authors report no conflicts of interest in this work.

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