

CASE REPORT

# Anesthesia Management in Scoliosis Surgery of Patients with Friedreich's Ataxia: A Report of Four Cases

## Friedreich Ataksisi Olan Hastaların Skolyoz Cerrahisinde Anestezi Yönetimi: Dört Olgu Sunumu

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ABSTRACT

Friedreich's ataxia (FRDA) is an autosomal recessive neurodegenerative disease, which may be accompanied by scoliosis, cardiac, endocrine and pulmonary comorbidities. In this paper the anesthesia experiences are presented using the total intravenous anesthesia (TIVA) method in scoliosis surgery of four patients with Friedreich ataxia. The patients were monitored with central venous pressure (CVP), bispectral index (BIS), invasive blood pressure, and near infrared spectroscopy (NIRS). The risks of difficult airway and malignant hyperthermia increased in patients with Friedreich's ataxia. Specialized equipment for difficult airway management was prepared and dantrolene was administered preoperatively. Neuromuscular blocker was used for anesthesia induction in one case, but was not required in the other 3 patients. Hypertrophic cardiomyopathy may accompany patients with Friedreich's ataxia. Therefore, detailed preoperative examination, intraoperative close monitoring of bleeding, blood gas analysis, hemodynamic monitoring and postoperative multimodal analgesia for hemodynamic stability were applied for patients with Friedreich's ataxia. The aim of this paper was to present our anesthesia management experience in four patients with Friedreich's ataxia who underwent scoliosis surgery, guided by the medical literature.

**Keywords:** Anesthesia; Scoliosis; Friedreich's Ataxia

ÖZ

Friedreich ataksisi (FRDA), kardiyak, endokrin ve pulmoner komorbiditelerin yanı sıra skolyozun da eşlik edebileceği otozomal resesif geçişli nörodejeneratif bir hastalıktır. Bu raporda Friedreich ataksili dört hastada, skolyoz cerrahisinde total intravenöz anestezi (TIVA) yöntemi ile anestezi deneyimlerimizi sunmayı amaçladık. Hastalar santral venöz basınç (CVP), bispektral indeks (BIS), invaziv kan basıncı ve near infrared spectroscopy (NIRS) ile izlendi. Friedreich ataksisi olan hastalarda zor hava yolu ve malign hipertermi riski yüksektir. Bu nedenle zor hava yolu için gerekli ekipman hazırlığı ve malign hipertermi riski için dantrolen sağladık. Bir olguda anestezi induksiyonunda kas gevşemesi sağlayabilmek için kas gevşetici kullandık, diğerlerinde kas gevşetici gerekmedi. Friedreich ataksisi olan hastalarda kalpte hipertrofik kardiyomiopati siktir. Bu nedenle Friedreich ataksili hastalarda hemodinamik stabilite için detaylı preoperatif değerlendirme, intraoperatif yakın kanama takibi, kan gazı analizi, hemodinamik izleme ve postoperatif multimodal analjezi önemlidir. Tıbbi literatür rehberliğinde, skolyoz cerrahisi geçiren Friedreich ataksisi tanısı olan dört hastadaki anestezi yönetimi deneyimimizi sunmayı amaçladık.

**Anahtar Kelimeler:** Anestezi; Skolyoz; Friedreich Ataksisi

Introduction

FRDA is an autosomal recessive and progressive neurodegenerative disease, with a reported incidence of approximately 1/50000. There is both dorsal root and ganglion degeneration in the spinal cord in FRDA patients (1). The disease is characterized by neurological symptoms such as progressive ataxia, dysarthria, areflexia in the lower extremities, muscle weakness in the legs, and non-neurological symptoms such as scoliosis, hypertrophic cardiomyopathy and diabetes mellitus (2).

In 95% of the patients, there is a GAA trinucleotide repeat increase in the first intron of the FRDA gene located on chromosome 9q13. This leads to a decrease in the level of frataxin protein, which has an important role in iron homeostasis. Iron accumulation in mitochondria and specific mitochondrial enzyme deficiencies caused by this, increased susceptibility to

oxidative stress and free radical mediated cell death are responsible for the pathogenesis of Friedreich's ataxia (3). There is no specific reference about the use of muscle relaxants in anesthesia management for scoliosis surgery of patients with FRDA. In these patients, the use of succinylcholine, a depolarizing muscle relaxant, is avoided due to the possibility of a hyperkalemic response through potassium release from denervated muscle cells (4).

In FRDA patients, progressive vertebrae curvature causes limitations in cardiac and pulmonary functions. Cardiac deterioration with left ventricular malfunction is common, and the quality of life decreases with the accompanying pulmonary dysfunction. The Cobb angle shows the level of vertebral curvature, and surgical intervention may be required in patients with a COBB angle higher than 40° (5). FRDA patients undergoing

scoliosis surgery require extensive preoperative evaluation and planning in terms of both surgery and anesthesia. Especially during anesthesia, accurate evaluation of neuromuscular block, avoidance of risks associated with hyperkalemia, and monitoring including intraoperative fluid balance are important. Appropriate management of postoperative pain also helps to prevent complications. Bleeding is another risk factor in patients undergoing long segment spine surgery and requires close intraoperative follow-up. The aim of this paper was to present the perioperative anesthetic management in four patients with FRDA who underwent long segment thoracolumbar biconcave scoliosis correction surgery.

### Case 1

Scoliosis correction surgery was planned for an 18-year-old male patient who was diagnosed with FRDA three years previously from the findings of tremor in the hands, balance disorder, chest pain and increased Cobb angle (51o). Physical examination revealed an atrophic appearance of all four extremities and neurologically omnidirectional horizontal nystagmus. In addition, cardiac examination of the patient revealed concentric hypertrophy in the left ventricle on echocardiography (ECHO) and repolarization abnormality in V1-V6 on electrocardiogram (ECG). There were no abnormal breathing sounds in the examination or signs of infiltration on the chest X-ray.

Dantrolene was prepared for the risk of malignant hyperthermia. For the difficult airway, the necessary equipment was provided preoperatively. The patient's Mallampati score was 2, and the ASA score was 2. The thyro-mental distance and mouth opening of the patient were within normal limits. Following routine anesthetic monitoring, preoxygenation was performed and endotracheal intubation was performed after anesthesia induction without the use of muscle relaxant. Invasive blood pressure monitoring, jugular central venous catheterization, BIS, and NIRS monitoring were performed. Anesthesia was maintained with total intravenous anesthesia. After basal values were obtained with somatosensorial evoked potential (SEP) and motor evoked potential (MEP), these were followed up during surgery. Tranexamic acid 10 mg/kg iv bolus and 1 mg/kg infusion were started as antifibrinolytics and were continued throughout the surgery. Arterial blood gases, electrolyte values, amount of bleeding, hemoglobin values and hourly diuresis were monitored. The mean arterial pressure was maintained at 60 mm Hg, and the BIS value was in the range of 30-60. NIRS decreased no more than 10-15% from baseline values.

As the surgical intervention, 22 transpedicular screws were placed between the T4-L3 vertebrae. Posterior stabilization was achieved with a rod placed on the right and left sides. Scoliosis correction was performed by distraction and compression. Posterior osteotomy was performed between T5-L2 and posterolateral fusion was applied to T5-L2. While the

initial hemoglobin value was 15 mg/dL, it was 10 mg/dL at the end of the operation, but there was no need for blood or blood product transfusion throughout the operation. During the surgery, blood pressure, pulse and NIRS values remained within normal limits. There was no change in the control neuromonitorization values of the patient compared to the baseline. The patient was extubated at the end of the operation and taken to the neurosurgery intensive care unit. Pain control was provided with local anesthetic infiltration, paracetamol, dexketoprofen and opioid analgesics if needed. The patient was transferred to the neurosurgery ward on the second postoperative day and did not experience any problems in the follow-up period. The patient was discharged on the 7th postoperative day.

### Case 2

A 15-year-old male patient complaining of unsteady gait 8 years previously was diagnosed with FRDA, familial mediterranean fever (FMF) and hypertrophic cardiomyopathy (HCMP). As a result of inability to walk without support, chest pain and increased Cobb angle (50o), scoliosis surgery was planned. In the preoperative physical examination of the patient, dysarthric speech, dysmetria, and dysdiadokinesia were present, and upper and lower extremity muscle strength was evaluated as 3/5. Cardiac examination revealed HCMP, mitral regurgitation (MR grade 1-2) on ECHO, T negativity in D2-3 AVF and V5-6 on ECG. Breath sounds were normal during inspiration, and no signs of infiltration were observed on the chest X-ray.

For prophylaxis of infective endocarditis, 2 gr ampicillin iv was administered preoperatively. The patient's Mallampati score was 3 and ASA was 4. The thyromental distance and mouth opening of the patient were normal. Following routine anesthetic monitoring, preoxygenation was performed with 100% oxygen for 3 minutes and induction of anesthesia with 30 mcg/kg midazolam, 1 mg/kg lidocaine, 2 mg/kg propofol, and 2 mcg/kg fentanyl, without using muscle relaxant. Endotracheal intubation was performed and the patient was ventilated with 7 ml/kg tidal volume. Invasive blood pressure monitoring was performed from the right radial artery, CVP monitoring with right subclavian venous cannulation, and BIS and NIRS monitoring. Anesthesia was maintained with TIVA. After basal values were obtained with SEP and MEP, they were followed up neurologically throughout the surgery. Tranexamic acid 10 mg/kg bolus and 1 mg/kg infusion were started as antifibrinolytics, and 1 mg/kg lidocaine was continued throughout the surgery. Arterial blood gases, electrolyte levels, amount of bleeding, hemoglobin values and hourly diuresis were monitored. The mean arterial pressure was measured as 65 mm Hg, and the BIS value was between 30-60. NIRS decreased no more than 10-15% from baseline values. As the surgical intervention, 26 transpedicular screws were placed between the T3-L3 vertebrae. Posterolateral stabilization was achieved by establishing the system with one rod each placed

on the left and right sides. Scoliosis correction was achieved by distraction and compression. The bones taken from the patient and 60cc bone chips were laid at the T3-L3 distance and posterolateral fusion was achieved. The initial Hb value was 13.7 mg/dL, and 10 mg/dL at the end of the operation, with no requirement for any blood or blood product transfusion throughout the operation. The control neuromonitorization values of the patient showed no change compared to the baseline values. After extubation at the end of the operation, the patient was taken to the neurosurgery intensive care unit. Local anesthetic infiltration was made for analgesia, and pain control was provided with paracetamol, diclofenac and tramadol. The patient was transferred to the ward on the second postoperative day, and with no problems experienced during follow-up, he was discharged on the 7th postoperative day.

### Case 3

A 24-year-old female patient was diagnosed with Friedreich's Ataxia 5 years previously. In the follow-up, scoliosis correction surgery was planned as a result of inability to walk without support, balance disorder and increased Cobb angle (41°). On physical examination, cerebellar tests were bilaterally clumsy, bilateral dysmetria and truncal ataxia were present, and speech was slowed with normal content. Heart rate was 89 bpm with sinus rhythm on the ECG. The pulmonary examination was within normal limits.

The patient's Mallampati score was determined as 2 and ASA was 2. The thyro-mental distance and mouth opening of the patient were normal. Following routine anesthetic monitoring, preoxygenation was performed with 100% oxygen for 3 minutes, then endotracheal intubation was performed after anesthesia induction with 1mcg/kg fentanyl, 3mg/kg propofol and 2mg/kg lidocaine without the use of muscle relaxant and the patient was ventilated with 7 ml/kg tidal volume. Invasive blood pressure monitoring was performed with right radial artery cannulation, CVP monitoring with right subclavian venous catheterization, and BIS and NIRS monitoring were performed. Anesthesia was maintained with TIVA. After basal values were obtained with SEP and MEP, these were followed up neurologically throughout the surgery. Tranexamic acid 10mg/kg bolus and 1mg/kg infusion were started as antifibrinolytics, and 1mg/kg lidocaine was continued throughout the surgery. Arterial blood gases, electrolyte levels, amount of bleeding, hemoglobin values and hourly diuresis were monitored. The mean arterial pressure was measured as 65-70 mm Hg, and the BIS value was in the range of 30-50. NIRS decreased by a maximum of 5-10% compared to baseline values. As the surgical intervention, 22 transpedicular screws were placed between T3-L3 vertebrae. Posterior stabilization was achieved by establishing the system with 2 rods. Scoliosis correction was achieved with distraction and compression. Posterolateral fusion was achieved by laying the bones taken from the patient and 60cc bone chips at the T3-L3 distance.

The initial Hb value was 14.8 mg/dL, surgical bleeding was monitored, and intraoperatively 1U erythrocyte suspension was replaced, and the hemoglobin value was 11.7 mg/dL at the end of the operation. There was no change in the control neuromonitorization values of the patient compared to the baseline. After extubation at the end of the operation, the patient was taken to the neurosurgery intensive care unit. Local anesthetic infiltration was performed for analgesia and pain control was achieved with dexketoprofen. The patient was transferred to the ward on the second postoperative day, and with no problems in the follow-up, she was discharged on the 5th postoperative day.

### Case 4

A 17-year-old male patient complaining of fatigue and muscle weakness was diagnosed with FRDA 6 years previously. As a result of the increase in Cobb angle (59°), scoliosis surgery was planned. The preoperative physical examination of the patient revealed that cerebellar tests were skillful, whereas deep tendon reflexes were hypoactive in the upper extremity, and deep tendon reflexes were absent in the lower extremities. Cardiac examination revealed mild hypertrophic changes in the left ventricle on ECHO and negative T in D2-3 AVF on ECG, therefore the troponin level was checked and was found to be within normal range. Breath sounds were normal during inspiration, and no signs of infiltration were observed on the chest X-ray. For the prophylaxis of preoperative infective endocarditis, 2gr ampicillin was administered to the patient. The Mallampati score of the patient was 2, ASA was 2, and the thyro-mental distance and mouth opening were normal. Following routine anesthetic monitoring, preoxygenation was performed with 100% oxygen for 3 minutes. Anesthesia induction was performed using 1 mg/kg lidocaine, 3mg/kg propofol, 2 mcg/kg fentanyl and 0.6mg/kg rocuronium. Endotracheal intubation was performed without any problems and the patient was ventilated with 8 ml/kg tidal volume. Invasive blood pressure monitoring was performed with right radial artery cannulation, CVP monitoring with right subclavian vein catheterization, and BIS and NIRS monitoring were performed. Anesthesia was maintained with TIVA. After basal values were obtained with SEP and MEP, these were followed up neurologically throughout the surgery. Tranexamic acid 10mg/kg bolus and 1 mg/kg infusion were started, and 1mg/kg lidocaine was continued throughout the surgery. Arterial blood gases, electrolyte levels, bleeding amount, hemoglobin values and hourly diuresis were monitored. The mean arterial pressure was measured as 70 mm Hg, and the BIS value was in the range of 30-60. NIRS decreased by no more than 10-15% from baseline values. As the surgical intervention, 24 transpedicular screws were placed between the T2-L2 vertebrae. Posterolateral stabilization was achieved by establishing the system with one rod placed on the left and right sides. Scoliosis correction was achieved by distraction and compression. The bones taken from

the patient and 60cc bone chips were laid at the T2-L2 distance and posterolateral fusion was achieved. The initial Hb value was 15.7 mg/dL, and was 9.8 mg/dL at the end of the operation, and 1U of erythrocyte suspension was replaced during the operation. There was no change in the control neuromonitorization values of the patient compared to the baseline. After extubation at the end of the operation, the patient was taken to the neurosurgery intensive care unit. Local anesthetic infiltration was performed for analgesia and pain control was achieved with paracetamol, dexketoprofen, and tramadol. The patient was transferred to the ward on the second postoperative day, and with no problems experienced during follow-up, he was discharged on the 4th postoperative day.

### Discussion

Uneventful anesthesia management was provided for four patients with Friedreich's Ataxia who underwent scoliosis surgery, with the use of appropriate hemodynamic monitoring and neuromuscular monitoring, total intravenous anesthesia, intraoperative close patient follow-up and a multimodal analgesia approach.

Patients with FRDA usually have bulbar symptoms, and are therefore at the risk of perioperative and postoperative aspiration. In addition, respiratory functions are limited due to thoracic kyphoscoliosis, which increases the risk of aspiration (6). There is a significant increase in risk for cardiomyopathy and congestive heart failure in patients with FRDA. One third of patients have impaired glucose tolerance or diabetes mellitus. Preoperative evaluation should be performed carefully because of neurological, cardiac, pulmonary and endocrine dysfunction (6,7). Precautions should be taken for difficult airway management secondary to kyphoscoliosis and comorbidities in this patient group (8).

The increased risk of malignant hyperthermia in patients with neuromuscular disease requires avoidance of volatile anesthetics and succinylcholine as much as possible (9). Schmitt et al. applied TIVA with propofol and sufentanil instead of volatile anesthetic to maintain anesthesia in two patients diagnosed with FRDA and reported that they did not observe any delay in recovery from rocuronium concluding that non-depolarizing muscle relaxants could be used safely in FRDA patients (8). Pancora et al. reported that the intubation of FRDA patients was facilitated with the use of propofol and sufentanil without muscle relaxants, when optimal drug infusion (remifentanil and propofol) was used to keep the BIS value between 45-60 during maintenance. Consequently, Pancora et al. did not experience any problems regarding hemodynamics during the operation (10). With the exception of one of the current cases, muscle relaxants were not used in the induction and maintenance of anesthesia.

### Conclusion

Detailed preoperative evaluation is important in anesthesia management of FRDA patients due to cardiac, pulmonary, neurological and endocrine comorbidities (9,10). Preparations for difficult intubation should be made due to kyphoscoliosis and accompanying comorbidities(7). Dantrolene should be prepared for the risk of malignant hyperthermia, although inhalation anesthetics and depolarizing muscle relaxants should be avoided. With TIVA, hemodynamic stability and intraoperative neurological evaluation can be provided during endotracheal intubation, throughout surgery and during recovery from anesthesia. Due to accompanying cardiac comorbidities in the FRDA patient group, the amount of bleeding should be closely monitored and postoperatively a multimodal analgesia method should be applied.

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