Anesthetic considerations in scoliosis patient with dopa-responsive dystonia or Segawa’s syndrome: Two case reports and a review of the literature

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Abstract
Segawa’s syndrome or dopa-responsive dystonia is a rare hereditary disorder characterized by progressive dystonia of childhood onset, diurnal fluctuation of symptoms and complete or near complete alleviation of symptoms with administration of low-dose oral levodopa. From our literature search in PubMed, we found only three related publications: two on anesthesia for cesarean section and one on anesthesia for electroconvulsive therapy. We report our experience in providing anesthesia for corrective scoliosis surgery in two biological sisters with Segawa’s syndrome. A review of the literature is also included.

Keywords
anesthetic, dopa-responsive dystonia, scoliosis, Segawa’s syndrome

Introduction
Segawa’s syndrome or dopa-responsive dystonia (DRD) is a rare hereditary disorder with an estimated prevalence of 0.5 per million population. Clinically, it is characterized by three hallmark signs: presentation of progressive dystonia of childhood onset, diurnal fluctuation of symptoms and complete or near complete alleviation of symptoms with administration of low-dose oral levodopa.

Given the rarity of DRD, we only found three publications on PubMed: two on anesthesia for cesarean section and one for electroconvulsive therapy. Here we report our experience in providing anesthesia for scoliosis surgery in two sisters. Permission to publish was obtained from the children’s parents.

Case reports
Case 1
A 14-year-old, 44-kg girl with DRD and neuromuscular scoliosis presented to our center for posterior spinal fusion. Since 7 years of age, she started experiencing generalized muscle weakness, clumsiness, and involuntary dystonic movements at rest. Her symptoms have a diurnal fluctuation. Metabolic, connective tissue and inborn error of metabolism screening, magnetic resonance imaging of brain, nerve conduction study, and muscle biopsy were done but unremarkable. At the age of 12, the diagnosis of DRD was made after a positive phenylalanine-loading test. She had a complete resolution of muscle weakness and dystonic movement after commencement of low-dose oral levodopa, 31.25 mg twice daily. Her thoracic scoliosis has a Cobb angle of 20° (Figure 1) and posterior fusion from T2 to T12 was planned. Preoperatively, she was asymptomatic and physical examination was unremarkable. Lung function

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