# Sevoflurane and thoracic epidural anesthesia for trans-sternal thymectomy in a child with juvenile myasthenia gravis

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#### Abstract

Literature on anesthetic management of juvenile myasthenia gravis (JMG) for thymectomy is limited. Recently, use of inhalational agents and total intravenous anesthesia with propofol and remifentanyl has been reported. All these techniques individually or in combination have been tried to avoid the use of muscle relaxant. We report successful use of sevoflurane as sole anesthetic agent for intubation and in combination with thoracic epidural anesthesia for intraoperative anesthetic management in a 5-year-old child with JMG.

Key words: JMG, sevoflurane, thoracic epidural anesthesia

#### Introduction

Myasthenia gravis (MG) is an autoimmune disorder characterized by weakness and fatigability of skeletal muscles. Children are affected in 10%-20% of MG cases and JMG is the most common type. However, 60%-90% children show improvement following thymectomy.<sup>[1,2]</sup> Available literature on the anesthetic management of JMG is limited and mainly consists of various techniques aimed at avoiding muscle relaxant.<sup>[1]</sup> We report a child with JMG after obtaining consent from parents who successfully underwent transsternal thymectomy under sevoflurane and thoracic epidural anesthesia.

### **Case Report**

A 5-year-old male child weighing 15 kg presented with bilateral ptosis for last 2 years and gradually progressive generalized muscle weakness for last 1 year which increased by the end of the day. Tensilon test was suggestive of MG

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and acetylcholine receptor antibody (anti-AChR) test was positive. However, he did not produce any history of recurrent respiratory tract infection, aspiration, or any other symptoms suggestive of bulbar muscle weakness. Subsequent contrast-enhanced computed tomography (CECT) chest revealed homogenous enlargement in the anterior mediastinum suggestive of hypertrophied thymus. Medical therapy in the form of oral pyridostigmine 15 mg, four times a day, prednisolone 20 mg once daily was initiated. The child had good compliance to medication. His generalized weakness improved, but ocular symptoms persisted even after 6 months and he was scheduled for thymectomy.

During preanesthetic check up, he was playful child without any bulbar weakness and normal muscle power 5/5. Hematology and blood biochemistry were normal. Chest x-ray showed mediastinal widening without any other abnormalities. CECT chest did not reveal any tracheobronchial narrowing, mediastinal or hilar lymphadenopathy, or involvement of mediastinal vessels, heart and esophagus.

The child was kept nil per oral for 8 h for solids and 2 h for clear fluids. Tab pyridostigmine and prednisolone were continued on the morning of surgery. In the operating room, oxygen saturation  $(SpO_2)$ , electrocardiography, and noninvasive blood pressure were attached and anesthesia was induced with sevoflurane 8% in oxygen. Neuromuscular monitoring (train of four-TOF) was attached and intravenous access was secured. After the child was apneic positive pressure mask ventilation was initiated. His TOF count was 4 and TOF ratio was 40%-50% at that time. Under deep

sevoflurane anesthesia (end tidal sevoflurane;  $E_{t_{sev}} 6\%$ ) direct laryngoscopy was done, larynx and trachea were sprayed with topical 10% lignocaine and trachea was intubated with 5.5 mm uncuffed endotracheal tube.

Epidural catheter (18G) was inserted at L<sub>2,3</sub> interspace and was advanced under ultrasound-guided hydrodissection technique till  $T_{\varepsilon}$  level. The catheter tip location was confirmed by epidurography using fluoroscopy. Epidural infusion of ropivacaine 0.2% with fentanyl 2 mcg/mL was initiated at 4 mL/h after initial 5 mL bolus. Anesthesia was maintained with  $O_2$ , air, and sevoflurane (Et<sub>sev</sub> 3%). Left radial artery was cannulated for invasive arterial monitoring. Midline sternotomy and thymectomy was performed. Surgery lasted for 2 h. Blood loss was around 70 ml and 400 mL of ringer's lactate was infused. His total urine output was 60 mL. TOF ratio throughout surgery was 20%-40%. No supplemental intravenous opioid analgesia was required. Tab pyridostigmine 15 mg was given through nasogastric tube as nearly 6 h had elapsed since last dose. At the end of surgery, sevoflurane was discontinued and trachea was extubated after clinical signs of adequate neuromuscular recovery were achieved and the child was fully awake. TOF ratio at that time was 80%. He was shifted to paediatric high dependency unit. Epidural ropivacaine 0.1% with fentanyl 2 mcg/mL was continued at 3-4 mL/h for 72 h. Tab pyridostigmine and prednisolone were continued as per schedule. He was discharged home after 7 days.

#### Discussion

JMG is associated with ocular symptoms like ptosis, ophthalmoplegia, strabismus, and in severe forms with generalized muscle weakness involving bulbar (dysphonia, dysphagia) and proximal limb muscles.<sup>[3]</sup> They are at risk of choking, aspiration, and chest infection.<sup>[2]</sup> Diagnosis of JMG is usually clinical with fluctuating weakness and fatigability. Further testing includes edrophonium test, repetitive nerve stimulation, and single fibre electromyography.

Acetylcholinesterase inhibitors are the first-line treatment. Pyridostigmine is commonly used in view of its oral tolerance, few muscarinic side effects, and long duration of action. Neostigmine may be used perioperatively in patients who are unable to take orally; or pyridostigmine may be continued through nasogastric tube as in the present case. Pyridostigmine 30 mg orally is equivalent to neostigmine 1 mg intravenous (IV)/Intramuscular (IM).<sup>[4]</sup> High dose steroids and immunosuppressant drugs like cyclosporine, azathioprine are used in progressive disease. Plasmapheresis and intravenous immunoglobulin (IVIg) are used during exacerbations and myasthenic crisis.<sup>[4]</sup> No specific anesthesia technique has been proven to be superior for thymectomy.<sup>[1]</sup> Use of muscle relaxant remains the most important consideration. Succinylcholine dose requirement is increased 3-4 times<sup>[3]</sup> and phase 2 blockade can occur since the threshold for causing depolarising block is exceeded. Hence, it should better be avoided.<sup>[3,5]</sup> Sensitivity to nondepolarizing muscle relaxants is increased as the amount of receptors needed for competitive antagonism is less and so more amount of drug remains at the junction than required.<sup>[3]</sup> Atracurium at a dose of 10%-20% of the normal dose has been suggested as the relaxant of choice.<sup>[5,6]</sup> Children on high dose steroids will require perioperative steroid supplementation.<sup>[7]</sup>

Current literature describes both total intravenous anesthesia (TIVA) and inhalational techniques to avoid/minimize the use of muscle relaxants. Sevoflurane has been found to provide adequate intubating conditions without any muscle relaxant It has been used as a sole anesthetic agent for trans-sternal thymectomy.<sup>[8]</sup> In a series of 68 adult patients for trans-sternal thymectomy, propofol, or sevoflurane anesthesia could avoid muscle relaxant and all the patients could be extubated in the operating room.<sup>[9]</sup> TIVA with propofol and remifentanyl infusion has been used for thymectomy which decreased the need for muscle relaxants.<sup>[1,3]</sup> Opioids at therapeutic concentration have no effect on neuromuscular transmission but central respiratory depression is a concern.<sup>[8]</sup> Remifentanyl has been found to provide an adequate depth for intubation with good recovery and can be used as an adjunct when muscle relaxant is not used.<sup>[1,3]</sup> However, there are reports of delayed postoperative recovery after remifentanyl infusion possibly due to inhibition of esterase degradation of remifentanyl by anticholinesterase.<sup>[10]</sup>

Epidural analgesia has been found to reduce the need for intraoperative and postoperative opioids and promotes early extubation.<sup>[1,3]</sup> However, direct thoracic epidural catheterization in small children may be risky. Hence, we chose lumbar route and advanced the catheter under real-time ultrasound guidance and confirmed the catheter placement under fluoroscopic guidance.<sup>[11]</sup>

Anticholinesterases used for reversal may show decreased response if the patient is already on pyridostigmine as per schedule. Moreover, it can precipitate cholinergic crisis.<sup>[3]</sup> Spontaneous recovery and extubation after assessment of bulbar and respiratory muscle function is preferred.<sup>[1,3]</sup> All patients are closely monitored in the postanesthesia care unit, where facilities for respiratory support are readily available.<sup>[3]</sup>

Thus to conclude, sevoflurane as a sole anesthetic agent provides good intubating condition and in combination with continuous thoracic epidural anesthesia provides adequate surgical anesthesia to avoid muscle relaxant in JMG.

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