Anesthetic Management in Pediatric Patient for Percutaneous Endoscopic Gastrostomy with Mitochondrial Myopathy: Leigh Syndrome

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Abstract
Leigh syndrome (LS) is a rare disease mainly affecting the central nervous system due to the abnormalities of mitochondrial energy generation and seen in early childhood with progressive loss of movement, mental abilities, seizures, nystagmus, ophthalmoparesis, optic atrophy, ataxia, dystonia, or respiratory failure. Anesthesia and surgery exacerbate the risks of aspiration, wheezing, and breathing difficulties. Tracheal irritability can be stimulated with the efforts of intubation. We report the anesthetic management of a rare case of an 11-year-old boy with a severe form of LS for percutaneous endoscopic gastrostomy insertion. The patient was closely monitored during the procedure and the postoperative period. Carefully chosen anesthetic agents, good pain control, and close monitoring are essential.

Keywords: Anesthesia, central nervous system, Leigh syndrome, rare

Introduction
Leigh syndrome (LS) is an extremely rare disorder with an estimated prevalence of 1 in 40,000 live births. It was first reported in 1951 by Denis Leigh.1 It is one of the member of the mitochondrial myopathies. More than 75 gene mutations, related to pyruvate metabolism and mitochondrial respiration, have been found responsible for LS. Brain changes are bilaterally symmetric lesions in thalamus, basal ganglia, and brain system due to the mitochondrial energy generation. It usually becomes apparent in the 1st year of life and rarely in late childhood and elderly years.2 A variety of clinical presentations can be presented. Thiamine infusion was given as supportive therapy.1,2

Clinical Presentation
The patient was an 11-year-old boy, weighing 30 kg, 110 cm height with second-degree consanguineous marriage admitted to our pediatric department for percutaneous endoscopic gastrostomy (PEG) insertion. He had an uneventful perinatal history and no family history of neurological disorders. He was diagnosed with LS at 2 years with status epilepticus and recurrent episodes of ataxia. Basal ganglia and cerebellar lesions were shown in brain magnetic resonance. His only medication included once-daily Keppra® (levetiracetam) for the recurrent seizures. On physical examination, he was mentally motor retarded, with ataxic gait. He had suffered from recurrent bronchopneumonia as a result of secretions and aspiration. He was dependent on nasogastric tube feeding. The patient needed oxygen and steam therapy daily at home. He had deformities of the thoracic bones and severe scoliosis, wheezing, and rough lung sounds which were detected during his physical examination and auscultation [Figures 1 and 2]. He also received muscle and joint physiotherapy for muscle spasms and spasticity.

Airway examination revealed a Mallampati score of III. His hematological workup and electrolytes were within normal limits.

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Written informed consent was obtained from his parents. He was transported to the gastroenterology unit where routine monitors were placed. An anesthesia machine, scavenging system, \( O_2 \) source, and routine monitoring were available. Sedation was accomplished with propofol intravenous (i.v.) 0.5 mg/kg maintained with a propofol 50–100 mg/kg/min and 0.5 \( \mu \)g/kg/min remifentanil i.v infusion. He received nasal oxygen breathing spontaneously. Oxygen saturation was maintained between 97% and 100%. Body temperature, heart rate, noninvasive blood pressure, \( O_2 \) saturation, and end-tidal \( CO_2 \) were stable during the PEG insertion and postoperative period. For postoperative analgesia, paracetamol 500 mg i.v. was given with no additional opioids. To prevent postoperative nausea and vomiting, granisetron 2 mg i.v. was administered preemptively. The procedure lasted 30 min and the total fluid administration included 250 ml of normal saline. He was discharged home on the same day.

**DISCUSSION**

Leigh disease has an approximately estimated prevalence of 1 in 40,000 live births. It usually presents in infancy with seizures, status epilepticus, developmental delay, dysarthria, and ataxia. These patients may also develop episodes of lactic acidosis that usually lead to respiratory failure and death.\(^3\) Anesthetic management of LS is often a challenge clinically. Typically, patients may require mechanical ventilation due to peripheral muscle weakness, multisystem organ failure, or respiratory insufficiency. Before surgery, anesthesiologists must keep in mind that the respiratory function impairment might progress in these patients. During surgery, all the general anesthetic agents are known to lead directly to perioperative problems.

Terkawi et al. reported a 15-year-old boy who underwent dental rehabilitation under general anesthesia. Anesthesia was induced with propofol and fentanyl. No volatile anesthesia was used. Propofol infusion was used during the surgery. Postoperative analgesia was managed with paracetamol and diclofenac sodium. No complications occurred, and the patient was discharged on postoperative day 2.\(^4\)

In another study Gozal performed, five children with LS underwent PEG insertion. Sedation was performed with propofol i.v. 0.5–1 mg/kg maintained with a propofol infusion 50–100 \( \mu \)g/kg and i.v. remifentanil infusion without complications.\(^5\)

Sasaki reported in 2008 a 17-year-old boy with LS undergoing laryngotracheal separation and open fundoplication. They used propofol and fentanyl infusion during the surgery.\(^6\)

In 2008, Footitt stated a review of patients with LS who underwent general anesthesia. In that review, postoperative respiratory failure and metabolic acidosis were reported in a 1-month-old patient, who underwent general anesthesia with propofol, sevoflurane, fentanyl, and midazolam.\(^7\)

Capnograph and pulse oximeter must be used routinely to detect hypoxia earlier in general anesthesia. Respiratory complications associated with the involvement of basal ganglia are frequent with LS. Volatile agents carry the risk of malignant hyperthermia, so should be avoided. We also did not use the long-lasting opioids because of the respiratory depressive effects. Another study performed by Jacobs et al. in 2004 reported the successful usage of remifentanil for scoliosis surgery in patients with LS as we used in our case.\(^8\) With the minimal respiratory depression effects and short duration of action, remifentanil is a good choice.

We suctioned the secretions during the whole procedure to minimize the obstruction of the airway. These children often have preexisting respiratory abnormalities and the secretions worsen the conditions.

There are a few reports in literature about LS and anesthetic management. Patients with myopathic conditions have longer recovery times after sedation or general anesthesia. Given its stable recovery profile, propofol and remifentanil can be good choice.

The disease is rare. Anesthesiologists should be aware of the respiratory function impairment. Further studies are needed.
to manage the perioperative period. With the increase of the cases in the future, it will become clear.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

**Conflicts of interest**

There are no conflicts of interest.

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