# Anesthetic concerns of Kleine–Levin syndrome

Dear Editor,

Kleine–Levin syndrome (KLS), an extremely rare (1–5 per million) autosomal dominant disorder, with male preponderance (M:F—2:1) and obscure etiology is characterized by acute episodes of frank hypersomnia, behavioral cognitive impairment, compulsive eating disorder, hypersexuality, and probable autonomic dysfunctions. These episodes last for 1–3 weeks followed by remissions of asymptomatic periods for 60–100 days which gets prolonged with disease progression.[1-3]

A 38-year-old female, BMI 36 kg/m$^2$, was scheduled for full-thickness corneal perforation repair following trauma, under general anesthesia. KLS was diagnosed at the age of 10 years, with symptoms persisting for 5 years and now being asymptomatic for last 22 years. Presently, she was on oral medications for psychiatric symptoms, hypothyroidism, and seizures (quetiapine, escitalopram, clonazepam, thyroxine, sodium valproate). She had infantile behavior requiring continual parental presence and assurance and was physically active by walking 30 mins every day. There were no gastro-esophageal reflux disease (GERD) and obstructive sleep apnea (OSA) (STOP-BANG-2/8), with Mallampati grade II and hyo-mental distance of 6 cm. Clinical examination and biochemical investigations including autonomic function tests were unremarkable.

On the day of surgery, all medications were continued. Her mother was allowed inside the operating room (OR) and her favorite song was played on the mobile. Difficult airway cart was kept ready. After attaching routine monitors including bispectral index (BIS), patient was placed on rapid airway management positioner (RAMP) [Figure 1], and anesthesia induction was done with titrated intravenous fentanyl, propofol, and atracurium and maintained with pro-seal laryngeal mask airway (PLMA) size 5 with air: oxygen mixture (1:1) and desflurane (minimum alveolar concentration 0.3–0.4 for BIS 40–60). Topical 0.5% proparacaine and intravenous paracetamol were given for analgesia. Surgery was completed uneventfully in 45 min. Neuromuscular blockade was reversed with neostigmine and glycopyrrolate. PLMA was removed when the patient was fully conscious. She was monitored overnight for any acute features of KLS and was discharged the next day. Patient is symptom free for last 4 months on telephonic follow up.

Preoperatively, stage of KLS should be determined as anesthetic consequences in acute phase may be grave. Triggering factors like infections, sleep deprivation, emotional stress, physical exhaustion, brain injury, and alcohol intake should be avoided.[1] Both general and local anesthesia may cause acute phase in KLS; however, there is no specific advice for any anesthetic drug. Thus, short-acting drugs were used. Postoperative monitoring and regular follow-ups should be done to detect reversion to acute phase.

Hyperphagia and increased leptin levels in KLS frequently result in obesity and OSA and thus hypoxemia, hypertension, atherosclerosis, GERD, and diabetes mellitus should be excluded. Invasive blood pressure monitoring maybe considered in major surgeries and in patients with autonomic dysfunction.[4] Drugs like stimulants, neuroleptics, mood stabilizers, and anti-convulsants have been tried in KLS and should be continued[1] [Table 1].

Other precautions taken were RAMP for difficult airway, continuation of medical drugs, monitoring of airway pressure for peri-operative seizures, and 2-min cycling of noninvasive blood pressure. To conclude, a gentle demeanor with a tailored approach of airway, endocrinopathies, and avoidance of triggering factors is crucial for safe anesthesia in KLS patients.

**Table 1: Salient features of Kleine-Levin syndrome and anesthetic concerns**

| Preoperative | Intraoperative | Postoperative |
|---|---|---|
| • Obesity<br>• Sleepdisordered breathing<br>• Endocrine disorders, for example, hypothyroidism<br>• Autonomic dysfunctions: hyperventilation, hypo/hypertension, Brady/tachycardia, thermoregulatory changes<br>• Meningeal symptoms: fever, headache, photophobia<br>• Behavioral disorder<br>• Treatment history | • Parental presence in OR<br>• Avoid triggers that may precipitate symptoms<br>• Increased sensitivity to respiratory depressants and opioids: apnea, prolong extubation time<br>• Short acting anesthetic agents<br>• BIS monitoring: maintain adequate depth, titrated anesthetic administration<br>• Neuromuscular monitoring<br>• Strict asepsis<br>• Avoid drugs lowering seizure threshold (i.e., sevoflurane, tramadol, tranexamic acid, certain antibiotics) | • Careful monitoring to detect respiratory depression<br>• Propped up position<br>• Followup to detect any acute episodes later |


**Figure 1:** RAMP position for expected difficult intubation

## Declaration of patient consent

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

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## Conflicts of interest

There are no conflicts of interest.

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## References

1. Arnulf I, Zeitzer JM, File J, Farber N, Mignot E. Kleine–Levin syndrome: A systematic review of 186 cases in the literature. Brain 2005;128:2763-76.
2. Mayer G, Leonhard E, Krieg J, Meier-Ewert K. Endocrinological and polysomnographic findings in Kleine-Levin syndrome: No evidence for hypothalamic and circadian dysfunction. Sleep 1998;121:278-84.
3. Miglis MG, Guilleminault C. Kleine-Levin syndrome: A review. Nat Sci Sleep 2014;6:19-26.
4. Freeman R, Chapleau MW. Testing the autonomic nervous system. Handb Clin Neurol 2013;115:115-36.



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